Splenosis

Ectopic location of the splenic tissue might proceed in the form of accessory spleen or splenosis. As distinguished from the inborn character of the accessory spleen, the presence of the splenic tissue is of intentional (controlled autotransplantation) or intrinsic, posttraumatic character (1, 2, 3). The phenomenon of the splenic tissue autotransplantation was first described in dogs by Graffini and Tizzani in 1883 and the name splenosis was introduced into modern terminology by Buchbinder and Lipkoff in 1939 (1, 2, 4, 5, 6). The initiating condition for the creation of splenosis is the splenic injury with capsule damage and the spread of splenic tissue. Zieliński and partners revealed the presence of the focuses of splenosis in 73% of the rats subjected to splenectomy and splenic tissue implantation into the peritoneal cavity (6). Healing of the splenic tissue fragments both in experimental and clinical studies was also described in Polish literature by Badowski, Orłowski and Ziarek (7, 8, 9). Intensification of the intrinsic implantation is ascribed to the degree of splenic fragmentation, the intensification of bleeding, the time period between the injury and its treatment, the local conditions and the use of intraoperative procedures, e.g. rinsing of the peritoneal cavity during the reconstruction (which limits the spread) and the frequency of the phenomenon is estimated at 67% (1, 6, 10, 11, 12).

The process of implants reconstruction proceeds most intensively during the first 5 weeks and depends on the level of their blood supply (1, 6, 13, 14). Reactivation of the implanted splenic fragments was described in isotopic tests in the period from 6 weeks to 24 months although there is information about contradictory results (7, 8, 15, 16). The most frequent location for autotransplantation is the peritoneal cavity. The focuses of splenosis were also described within kidney, liver, pancreas and paravertebral locations as well as in a laparotomy scar (2, 5, 17-22). The focuses of splenosis within solid organs, in soft tissues of the left arm and, especially, the intracranial focus of splenosis described by Rickert might prove the haematogenous character of the implants although it has not been confirmed unequivocally with experimental studies (23-26).

Thoracic splenosis

In ca. 18% of abdominal-thoracic injuries proceeding with spleen as well as diaphragm and lung damage, fragments of splenic tissue are implanted both within the peritoneal cavity as well as within the pleural cavity, the lung and the mediastinum (fig. 1) (5, 12, 27).
Thoracic splenosis

Left-sided abdominal-thoracic shots as well as traffic accidents are dominant in etiology. The first description of thoracic splenosis (TS) provided by Shaw and Shaff comes from 1937 (4, 28, 29, 30). Until 1995, 23 such cases were documented and their number is still growing. So far, over 70 cases of TS have been described with pulmonary-pleural location, most of which were left-sided. Most often, they were of asymptomatic character and were detected by accident as circumferential or parietal round shadows in the left lung with the diameter of 1-9 cm, and even 12 cm (most often up to 3 cm), or radiological pleural infiltrations. Other forms were also described imitating neuroblastomas, mediastinal and pericardial infiltrations as well as esophageal fibromas. In some of these cases, the reasons for hospitalization were febrile states and left-sided thoracic pains, even those of coronary type. A case was also described imitating lung carcinoma manifesting itself with recurrent hae-moptysis. The time period between the injury and the detection of the changes was within the range of several to 42 years. The macroscopic appearance of these changes had the form of soft, infiltrating or encapsulated, dark tumours in some cases taking on red, blue or purple colour, in 25% solitary and in 75% multiple, of various sizes. The changes were found on the parietal (most often) and visceral pleura, pericardium, in mediastinum, lobe fissures and lung parenchyma.

The patients were 15-79 years of age with the majority of men in accordance with the environmental risk. They did not usually exhibit any haematological and immunological disorders connected with the lack of spleen, i.e. asplenia. Due to the character of radiological changes, they were sent for diagnostics with suspected metastases or primary carcinoma manifesting itself with recurrent hae-moptysis. The result of the cytological examination of sputum, bronchial washings and specimens are negative in case of the changes located on the periphery of the lungs. Sonography controlled FNAB is preferred for the shadows adherent to the pleura (47). The changes located deeper in the lung parenchyma require a FNAB controlled with the X-ray or CT.

The lung tumours disqualified from FNAB are those located deeper than 7-8 cm from the skin level (needle range <11 cm), hilar tumours and those in patients without the required respiratory reserves in spirometry (FEV1 < 30%) or efficient haemostasis. The method sensitivity is estimated at over 90% and its specificity at over 73%. The method is not free from complications, the most frequent of which is pneumothorax and the most serious one is brain aeroembolism. The reduced FNAB diagnostic value is recorded in case of changes smaller than 1 cm and the most comfortable conditions for biopsy give tumours in the areas of lung segments II, VI and IX (44). However, in case of splenosis, and especially TS, the result of the fine-needle biopsy is ambiguous, difficult to evaluate and the interpretation of the results is different in the extreme. Galloro and Syed obtained lymphatic tissue as well as lymphocytes, macrophages and an outline of the tissue structure, which, together with the interview data, made the final diagnosis possible (17, 37). In most papers, however, the preparation was described to consist of mesenchymal cells as well as small and medium size lymphocytes or plasmatic cells partially without features of atypia and partially suspected of proliferating character. In at least 14 cases, the image of the FNAB preparation was simply non diagnostic (24, 25, 27, 29, 31, 32, 33, 35, 43, 45, 48-51). The use of the core biopsy or tru-cut biopsy enabled some authors to make precise diagnosis by obtaining a larger tissue fragment with the preserved splenic tissue structure (25, 52, 53, 54). In connection with the above, TS was most often recognised in the open biopsy of the changes through thoracotomy, minithoracotomy or videothoracoscopy (13, 14, 25, 28, 29, 42, 50, 52, 55). The presence of active, ectopic spleen focuses might be confirmed scintigraphically, also for the changes visible within the thorax. The used types of scintig-
raphy include scintigraphy with the use of sulphur colloids marked with Technetium-\textsuperscript{99m}Tc, scintigraphy with the use of one’s own thermally damaged erythrocytes as well as erythrocytes marked with \textsuperscript{99m}Tc and scintigraphy with the use of thrombocytes marked with Indium \textsuperscript{111}In. They differ in methodology, specificity and sensitivity. Scintigraphy with the use of sulphur colloid is the simplest, the cheapest and the most popular one although it is the least sensitive.

Scintigraphy with the use of one’s own erythrocytes and thrombocytes marks the spleen focuses selectively, is characterized by greater specificity, but is also more difficult technologically and more expensive. Scintigraphy with the use of one’s own erythrocytes is the most sensitive one, as it might be positive even in case of negative result of the scintigraphy with the use of sulphur colloid (15, 28, 30, 34, 39, 55-62). Overlapped isotopic images in versions SPECT and CT additionally increase the examination quality (63). Leucocytes marked with technetium were also used in isolated cases (59, 64). In one case, the diagnosis was based only on the CT examination (12). Such extended diagnostics made it possible to reduce the number of explorative thoracotomies.

Although Roucos described in 1990 that 13 out of 16 patients required open lung biopsy, currently this percentage is close to 50% (29, 65). Also, the number of cases diagnosed only with isotopic examination is increasing. Scales and Lee reported in 1983 that only 1 out of 9 cases of TS was diagnosed non-surgically, while currently the TS diagnosis has already been made in 19 cases based on scintigraphy (23, 24, 28, 30, 34, 39, 41, 57, 58, 60, 62, 64-69). An equally important diagnostic element is the evaluation of the cell elements of the circumferential blood, especially any possible abnormalities in the construction of the erythrocytes (Heinz bodies, Howell-Jolly bodies, shield-shaped blood cells, spherocytes, syderocytes and others) being the measure of the lacking spleen function. Vast majority of the evaluated patients did not exhibit any features of asplenia. The presence of Howell-Jolly bodies was recorded in only 2 patients and explained with the implants insufficiency (1, 23, 32, 43, 52, 55-70, 60, 65, 71, 72, 73). Based on the overview of the diagnostic methods, it might be concluded that the lack of improper forms of erythrocytes, the positive results of scintigraphy, the lymphatic tissue in the FNAB image and, most of all, the interview data about the abdominal-diaphragmatic-thoracic trauma with splenectomy are the key to diagnosing the active and functional focuses of TS.

Recently, the usefulness of MRI (ferromides-enhanced, SPIO-MRI) was described for the diagnostics of the focuses of TS with the use of iron oxides. A signal of a TS focus in phase T1 is distinctly strengthened after administering a Gadolinium bolus and the analogical signal of phase T2 is distinctly weakened after administering an iron oxide preparation. The image is identical for the spleen and the focuses of TS within the thorax and the phenomenon is explained with a specific capture of iron oxides e.g. in the reticular endothelium system, analogically to the capture of sulphur colloid in the isotopic examination. A case of TS diagnosed in this way has already been described (19, 31, 39, 48, 50, 59, 61, 68, 74).

Accessory spleens

The problem of differentiating the phenomena of splenosis and accessory spleens has been mentioned in the chosen articles (fig. 2) (12, 60). Al.-Ahmadi compiled such features as: frequency, etiology, number, location, blood supply and their size (tab. 1) (2). Differences in location, size, blood supply and structure, particularly the capsule structure, were described by Barcikowski and Harsh Mohan. The implants capsules do not contain any muscle or elastic fibres and their blood supply comes

![Fig. 2. Accessory spleen. Difference of view of vessels in comparison to the focus of splenosis](image-url)
Table 1. Comparison of main features of accessory spleens and focuses of splenosis

<table>
<thead>
<tr>
<th></th>
<th>Accessory spleen</th>
<th>Splenosis</th>
</tr>
</thead>
<tbody>
<tr>
<td>Incidence</td>
<td>10-44% of all necropsies</td>
<td>do 67%/ up to 67%</td>
</tr>
<tr>
<td>Etiology</td>
<td>congenital</td>
<td>acquired, history of splenic trauma, almost</td>
</tr>
<tr>
<td></td>
<td></td>
<td>invariably present</td>
</tr>
<tr>
<td>Number</td>
<td>1-6 / few, 1-6</td>
<td>numerous nodules – as many as 400</td>
</tr>
<tr>
<td>Location</td>
<td>okolica więzadła żołądkowo-śledzionowego /</td>
<td>any intra- or extraperitoneal location</td>
</tr>
<tr>
<td></td>
<td>near the gastroplenic ligament</td>
<td></td>
</tr>
<tr>
<td>Blood supply</td>
<td>wnękowe gałązki tętnicy śledzionowej /</td>
<td>newly formed arteries penetrating the</td>
</tr>
<tr>
<td></td>
<td>branches of splenic artery via the hilum</td>
<td>capsule</td>
</tr>
<tr>
<td>Size</td>
<td>may become as large as the original spleen</td>
<td>usually small, limited by the blood supply</td>
</tr>
</tbody>
</table>

from their environment penetrating through the capsule (1, 3). Rudowski, based on 611 splenectomies due to haematological reasons, specified the occurrence frequency of the accessory spleens at 18% in 99% cases in the number of 1-2. He also described other, potential (intra- and extraperitoneal) locations of the accessory spleens, however, outside the thorax (75, 76). Orlowski and Piecuch described the mechanisms for the creation of the accessory spleens emphasising, in order to differentiate both phenomena, the multiplicity and irregularity of the implants in posttraumatic cases (77). The increase in frequency of the occurrence of accessory spleens from 7.7% to 45% was described by Wkisbroth in his experimental study on rabbits with the use of phenylhydrazine, explaining the phenomenon as the answer of the organism to an increased demand for phagocytary efficiency, also in the cases after splenectomy (78). Such process might accompany a spleen damage caused by another pathological process, e.g. a splenic cyst (fig. 3).

Accessory spleens, usually singular, most often occur near the splenic hilum. They are blood-supplied through the branches of the splenic artery and retain the structure of the normal spleen (1, 75). The focuses of TS, multiple in 75%, are non-peduncular, blood-supplied through vascular branches of the parietal pleura, the pericardio-pleural artery, the diaphragm and the lung tissue. They are of various diameter although TS preparations were also described as mature splenic tissue (13, 14, 24, 32, 36, 37, 45, 48, 50, 51, 61, 65). While the presence of the accessory spleens in haematological affections might be the reason for the relapse of hypersplenism, the presence of the focuses of TS is connected with the return of the sequestrating functions of the splenic tissue (1, 13, 36, 75, 77).

Also, the regression of the focuses of TS was reported in case of the focuses in the peritoneal cavity taking over the functions (5). Balacumaraswami described the focuses of the accessory spleens (spleniculi) within the right pleural cavity in a patient with inborn spherocytosis and undergone, planned splenectomy several years earlier, with the CT image close to the image of the chosen focuses of TS (fig. 4) (79). In the available literature, the size and location of the focuses of TS and the accessory spleens within the thorax, thus, are not a decisive factor especially for the singular focuses of TS. In the context of radiological diagnostics of the thoracic tumorous shadows, it is difficult to make differentiation in the cases without any documented injury penetrating through the diaphragm.

Fig. 3. Splenic cyst in the CT view. There was not resected accessory spleen close to the splenic hilum revealed intraoperatively (data of the authors)
Consequences of splenectomy

The immunological and regulatory role of the spleen for the cellular blood elements is well recognised (15, 70, 71, 73, 76). The early consequences after the removal of the spleen due to an injury include e.g. leucocytosis, thrombocytosis, lymphocytosis, reticulocytosis and decrease in the level of antibodies of class IgM, however, the values of the above mentioned factors normalise within several months after the loss of spleen (15, 70, 71, 73, 76, 80).

The disappearance of the sequestering and filtrating-fagocytary functions results in the presence of the following new elements or varieties of typical cellular elements in the circumferential blood smear: Heinz bodies, Howell-Jolly bodies, syderocytes, spherocytes, shield-shaped blood cells and others (15, 43, 70, 71, 73, 76, 80, 81). The most important consequences of the loss of spleen include the decrease in the immunity to bacterial infections, mainly with capsular bacteria (Streptococcus pneumoniae, Haemophilus influenzae, Neisseria meningitidis) and viruses (15, 24, 67, 73, 76, 82, 83 – 86). The first observations on this subject conducted by Morris and Bullock in 1919 were confirmed by King and Shumacker in 1952 (11, 15, 76, 80, 81, 85, 86, 87).

Clinical meaning of the splenosis phenomenon, including the thoracic splenosis, involves the normalization of the circumferential blood smear in the further period after the splenectomy. The lack of any improper forms of erythrocytes and Howell-Jolly bodies corresponds to the fact that the focuses of splenosis take over this function. The described cases of TS, evaluated many years after the removal of the spleen, did not have the usual features of asplenia, but visible focuses of TS were a substitute form of the activity of the splenic tissue (15, 22, 24, 25, 28, 29, 31, 32, 41, 43, 48, 51, 55, 58, 62, 66, 68, 86).

Treatment procedure

Most indications for surgical treatment of the spleen are based on the resection procedures. In case of injuries, the indication for the removal of the spleen is its disaggregation with an uncontrollable haemorrhage. In many types of splenic injuries with smaller scale of trauma, it is possible to make use of the saving operations or even conservative treatment (7, 8, 10, 15, 16, 70, 71, 73, 76, 83, 86, 92, 93). Indications for planned splenectomy include a group of affections such as hypersplenism, spherocytosis, haemolytic anaemia, idiopathic thrombocytopenic purpura, chosen oncological cases, splenic vein thrombosis, Felty’s syndrome, splenic cysts and others (15, 71, 73, 76). An acknowledged protective method in case of the planned splenectomy, especially in groups of increased risk, i.e. children and adults >65, is vaccinations and chemoprophylaxis of infections (70, 71, 73, 76, 80, 82, 85, 93, 94).

Many works described the autotransplantation of the splenic fragments, most often within the greater omentum, in order to prevent the asplenia syndrome. In Polish literature, the technique for the procedure was provided by e.g. Orłowski, Badowski, Fibaik, Gajewski and Ziarek (1, 7, 8, 9, 16, 86, 89, 94).

Although the presence of the splenic autotransplants in the peritoneal cavity after the traumatic splenectomy is not a pathological symptom, other symptoms described for this reason...
including ileus, internal haemorrhage and acute abdominal pain, are the ground for the immediate laparotomy (10, 76, 95). In case of any recognisable focuses of splenosis being present in pulmonary-pleural locations, it is recommended to undertake saving procedures and avoid their removal (14, 19, 20, 22, 24, 28, 29, 31, 33, 48, 50, 55, 58, 60, 62, 65, 67, 68, 74).

Polish literature

There is vast Polish literature available on the spleen, the phenomena connected with its damage and their consequences. The basic subject was the removal of the spleen due to traumatic reasons (70, 92). A large number of clinical studies in the 1980s regarded the subject matter of the splenic tissue autotransplantation after an injury, both the intrinsic and the intentional, controlled one (1, 16, 86, 89, 94, 96). A model of controlled autotransplantation was described in the experimental studies of Zieliński, Badowski and Górecki (6, 90, 97). “Experimental and clinical studies on the implantation of the splenic parenchyma after splenectomy” was the subject of the habilitation dissertation of Badowski (15). Another group of works was devoted to the acute abdominal complications arisen on the basis of the intrinsic intraperitoneal autotransplantation as well as the subject matter of the accessory spleens, including their differentiation from the focuses of splenic implants (10, 75, 77, 96, 95). Also, the problem of splenic cysts, their etiology and treatment was taken up (83, 93). In 1982, a “Spleen surgery” monograph edited by Rudowski and Pawelski was published (76). The introduction of the laparoscopy technique into spleen surgery resulted in further reports on its use (98). An important group of papers is those on the results of spleenectomy and asplenia. The phenomena occurring after the removal of the spleen were the subject of case and clinical studies, congress reports and case descriptions (73, 80, 81, 84, 85, 88, 91). In spite of a wide spectrum of the quoted works, many other problems, elaborations and dissertations connected with spleen surgery were not mentioned due to editorial limitations. Although the subject of the post-traumatic autotransplantation of the splenic tissue was widely mentioned in the quoted material and Rudowski, Orlowski, Barcikowski, Zieliński and Badowski reported the possible locations of the intrinsic implants within the thorax, there has been no mention about TS encountered in the available Polish literature since 2006.

Polish report

The issues of the Polish Journal of Surgery 2006; 78: 1012-1016 and the Polish Pneumology and Allergology 2008; 76: 456-459 published the description of the case of autotransplants of the splenic tissue in the form of a tumour of the left lung in a 43 year old patient, who was subjected to laparotomy due to traumatic reasons, but did not have any documentation of previous treatment (45, 46). The circumferential shadow of the left lung was detected radiologically. From the operation until now, the course of the affection has been asymptomatic and the ground for the hospitalization was the prolonged bronchitis. The diagnostics made use of e.g. CT and sonography of the thorax and the abdominal cavity, bronchofiberoscopy and fine-needle biopsy (FNAB) of the round shadow in the left lung. A change with 3cm diameter was visible in the left lung together with several smaller nodules of the left-sided parietal pleura and some nodules in the left subdiaphragmatic area corresponding to the image of the metastatic carcinoma process as well as the lack of spleen (fig. 5, 6). The lung biopsy indicated the presence of the mesenchymal cells of medium size without any features of atypia. The pathologist suggested the necessity for an immediate ex-

Fig. 5. TS (arrow) as well, as coin lesion of the left lung of the polish case
amination from the open biopsy. The left-sided minithoracotomy indicated the diffuse pleural adhesions as well as more numerous, in comparison with the CT image, soft, black, infiltrating tumorous changes with the diameter of 0.5-3 cm in the pleura, the lung and the aortic-pulmonary window. The changes were most numerous in the area of the scar on the diaphragm after a suffered injury, which was revealed during the operation. All the changes reminded and mimicked the metastases of the malignant melanoma, as in other cases operated by the authors of this and other reports (12, 13, 26, 68).

In view of the spread process, only singular tissue fragments of the changes were taken and the procedure was finished. Histopathologically all the material was described as the autotransplants of the splenic tissue. In a later period, agreed with the patient, control examination was conducted as regards the activity of the splenic tissue. The patient was subjected to scintigraphy with the use of sulphur colloid marked with $^{99m}$Tc and numerous locations of the radioisotope accumulation were visible. These locations correlated with the CT image and corresponded to the focuses of splenosis within the peritoneal cavity as well as in the left lung with simultaneous lack of any features of asplenia (Howell-Jolly bodies index – 1/preparation, acanthocytes – 3-4/1000 erythrocytes, spherocytes sporadic/preparation, shield-shaped blood cells – 2/1000 erythrocytes, damaged blood cells – 2-3/1000 erythrocytes and the level of antibodies of class IgM – 1.95 g/l, norm 0.5-3.5 g/l (fig. 7). The other variants of the scintigraphy were not used due to technical problems. The interview and the course of the Polish case fits with the image described by the international literature. A serious difficulty for the initial evaluation was the lack of documentation from the treatment in the period of the suffered spleen injury together with the lack of any knowledge on the damage of the diaphragm. The radiological image, similarly to other reports, required differentiation from the metastatic carcinoma and the difficulties with the evaluation of the material from the FNAB, similarly to some reports, justified the qualification for the open biopsy of the lung. It is also important to add that in our case the videothoracoscopy would not have any use due to the diffuse pleural adhesions. The postoperative diagnostics of the Polish case did not involve the evaluation of the circumferential blood smear or any scintigraphic examinations concentrating on the potential, oncological aspect of the visible changes. Their use for such chosen cases in the future is a clinical recommendation.

Fig. 6. Focus of the intraperitoneal splenosis (arrow) in the upper left abdominal field of the polish case

Fig. 7. Numerous focuses of intensive concentration of technetium in the abdominal field (b) and above the left hemidiaphragm in the position of tumor of the left lung (a). Position of the liver (c)
CONCLUSION

TS is a rare complication of the acute abdominal-thoracic injuries with the undergone, documented splenectomies, which involve the development of the ectopic, implanted focuses of the splenic tissue with the potentially beneficial for the patient immunological, sequestrial and fagocytary activity. TS very often coexists with the focuses of splenosis within the peritoneal cavity (45, 46, 61, 62). Until recently the international literature has described over seventy such cases, almost always within the left side of the thorax. The reasons were either traffic accidents or shots. Changes of the type of the circumferential circular shadows or pleural-mediastinal infiltrations detected radiologically many years after the suffered injury had the etiology, which was difficult to specify and imitated the metastatic carcinoma. The reason for the hospitalization of the asymptomatic patients were the detected radiological changes and, for some patients, most often the unspecified pains in the left side of the thorax. In most cases, the oncological diagnostics focused on the FNAB and the open biopsy of the lung. However, if splenosis was suspected, the concentration was on the scintigraphic examinations and avoidance of the operative procedures, which is a "golden standard" in these patients (52, 54, 65, 72). It is also important to emphasise once again that the patient diagnosed because of the changes in the lungs, especially left-sided ones, with the undergone posttraumatic splenectomy, even with incomplete interview regarding the penetration of the injury through the diaphragm, must be suspected of the presence of the focuses of splenosis within the thorax. The diagnostic standard including sonography, CT, bronchofiberoscopy, fine-needle biopsy and, in some cases, PET must be supplemented with, at least, scintigraphy with the use of sulphur colloid marked with technetium$^{99m}$Tc and the circumferential blood smear, which, in case of the active focuses of TS, should not exhibit any features of asplenia. Probably, in the near future SPIO-MRI will become an equivalent diagnostic priority for the cases of TS in relation to the scintigraphic examinations.

A separate problem is the coexistence of carcinoma and other malignancies, especially lymphomas, and focuses of splenosis within the encountered changes in the lungs or pleural cavity already detected in 4 cases (12, 34, 37, 66, 69). It is also worth mentioning that the splenic angiosarcoma, with metastases to the peritoneal cavity and the lungs might imitate splenosis in its appearance (99). The decision about subjecting the patient to observation must, thus, take into account the complete evaluation of all the encountered changes. The lack of progress in the non-invasive diagnostics justifies the decision about the open biopsy of the lung although the use of videothoracoscopy is not always possible due to the previous type of injury penetrating into the pleura with a tendency for its adhesion (28, 48, 65).

The case described by the authors has, so far, been the first Polish report of splenosis with pulmonary-pleural location and the experience following from such unique clinical image should be helpful in diagnostics and treatment of the patients with round shadows in the lungs and pleural infiltrations in accordance with the following rule: „Consider the diagnosis of splenosis for soft tissue masses long after any splenic injury” and „If you suspect it, you can avoid thoracotomies” (13, 100).

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