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Myxoid liposarcoma with gastric localisation – Case Report

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ABSTRACT
Liposarcoma is the most common malignant mesenchymal neoplasm affecting soft tissues and, less frequently, viscera. Gastric localisation of liposarcoma is extremely rare. We present the case of a 69-year-old female patient, who underwent subtotal gastrectomy with processing of the specimen in the Clinical Service of Pathology, Emergency County Hospital of Constantza. Histopathologically, the tumor exhibits a neoplastic population composed predominantly of lipoblasts in different stages of differentiation, with a massive disposition involving submucosa, muscularis and serosa. These cells contain intracytoplasmic small optically empty lipid vacuoles that produce indentations of centrally located nuclei; isolated lipoblasts have the appearance of signet ring cells. Predominant cellular population is admixed with scattered mature adipocytes and with stellate/spindled mesenchymal cells. Tumoral stroma has a myxoid character. A distinctive feature of this neoplasm is the existence of a vascular network composed of curved thin-walled capillaries with a branching pattern ("chicken-wire"). The histopathological traits and immunohistochemical profile of this tumor, characterized by positive reaction to Vimentin and S100 and negative staining to pancytokeratin, support the diagnosis of myxoid liposarcoma. The rare incidence of this tumor in the stomach requires a careful microscopical examination, in order to differentiate it from other clinico-pathological entities and to apply an appropriate therapy with a proper monitoring protocol.

Keywords: myxoid liposarcoma, lipoblast, mesenchymal neoplasm.

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Introduction

Liposarcoma is the most common malignant mesenchymal neoplasm affecting soft tissues and, less frequently, viscera. Gastric localisation of liposarcoma is extremely rare [1], it involves mostly the lesser curvature and appears as a protrusive mass attached to the gastric wall. Microscopically, it is presumed that its origin lies in undifferentiated mesenchymal cells that proliferate within the submucosa and the muscularis propria of the stomach [2]. This exceptionally rare gastric malignant tumor must be distinguished from other non-epithelial and epithelial gastric neoplasms, in order to apply an appropriate therapy and a proper monitoring protocol [3].

Materials and methods

We present the case of a 69-year-old female patient, who was admitted to the Emergency County Hospital of Constantza due to inapetence, epigastric pain and gastric fullness. The patient has undergone an endoscopical examination that showed the presence of an exophytic lesion localised in the body of the stomach, apparently originating from submucosal layers and covered by normal mucosa. MRI procedure revealed a heterogeneous mass of the gastric wall composed of areas with features of adipose tissue admixed with foci of higher density,

as it was evidenced by intravenous administration of contrast substance. The patient underwent subtotal gastrectomy in the Surgery Department. The specimen was processed in the Clinical Service of Pathology, by opening on the great curvature and fixing in 10% formalin. Tissue blocks were sampled from all grossly different patterns of the tumor in order to examine them microscopically and to view the relationship between the neoplastic proliferation and the adjacent histologic structures. Sampled fragments were paraffin-embedded, sectioned at 5-µm and stained with Hematoxylin–Eosin (HE) and van Gieson. Immunohistochemical analysis was based on a panel of antibodies:

- Polyclonal rabbit anti-S100 (DAKO): cytoplasmic and nuclear positive reaction; helpful for the recognition of lipogenic tumors [4];
- Monoclonal mouse anti-vimentin, clone Vim 3B4, isotype IgG2a, kappa (DAKO); cytoplasmic reaction; marks cells of mesenchymal origin [4];
- Monoclonal Mouse Anti-Human Cytokeratin Clone AE1/AE3 – cytoplasmic immunostaining of all types of epithelia; supports or excludes the epithelial origin of tumors [4].

Results

The gross examination of the subtotal distal gastrectomy specimen of 10/7/1,5 cm reveals the presence on the lesser curvature of a protrusive submucosal and intramural nodular lesion with a diameter of 6/2,5 cm, relatively well defined, with a translucent multinodular yellowish appearance on section and a medium consistency (Figure 1).

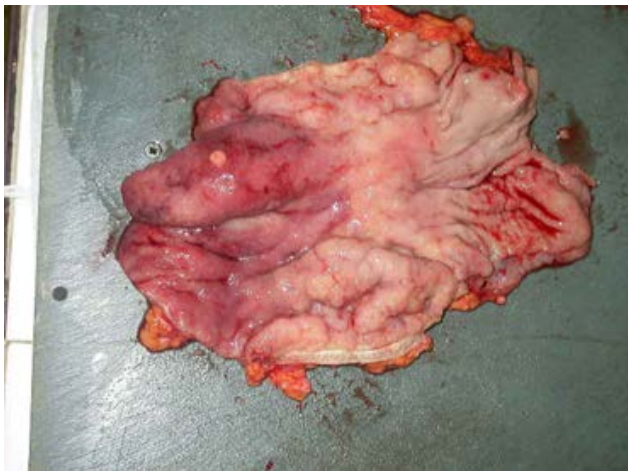

Figure 1. Gross appearance of gastric tumor

Overlying and adjacent gastric mucosa shows areas with thickened folds, alternating with zones of flattening. Histopathologically, the tumor exhibits a neoplastic population composed predominantly of lipoblasts in different stages of differentiation, with a massive disposition involving submucosa, muscularis and serosa (Figure 2).

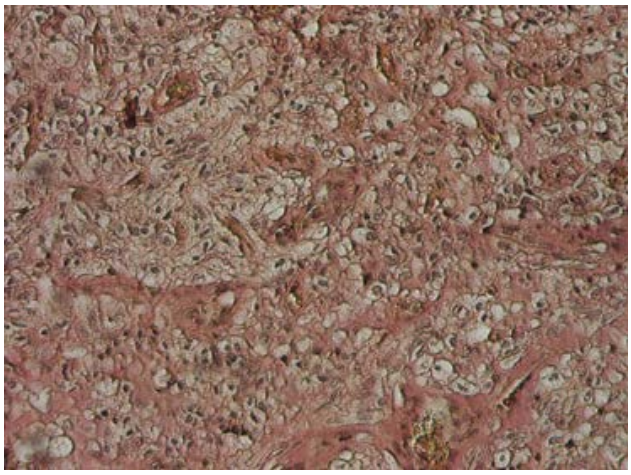

Figure 2. Neoplastic population composed predominantly of lipoblasts in different stages of differentiation (van Gieson X 100)

These cells contain intracytoplasmic small optically empty lipid vacuoles that produce indentations of centrally located nuclei; isolated lipoblasts have the appearance of signet ring cells (Figures 3 and 4).

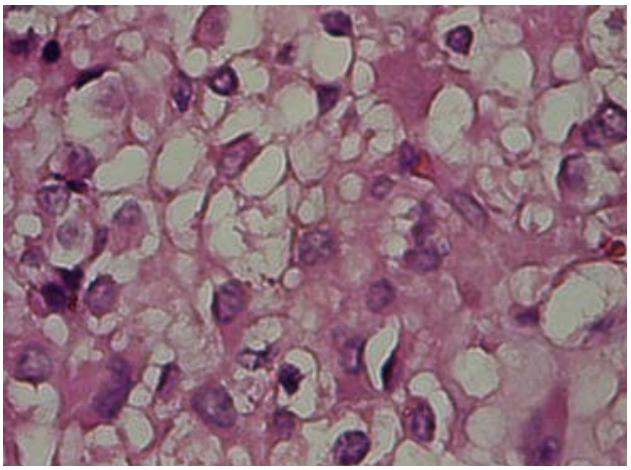

Figure 3. Lipoblasts with intracytoplasmic lipid vacuoles that produce indentations of centrally located nuclei (H.E. X400)

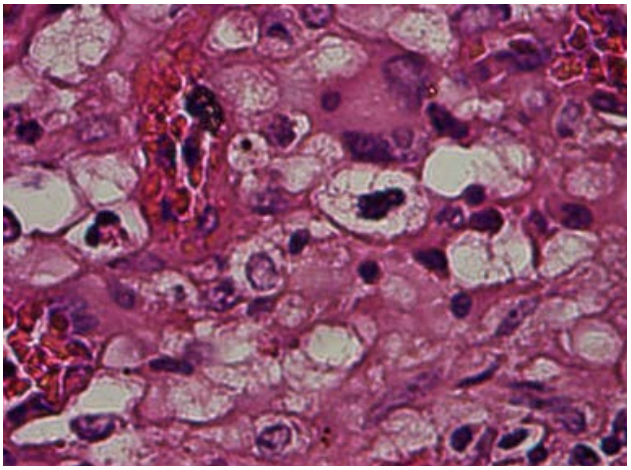

Figure 4. Lipoblasts admixed with signet ring cells (H.E. X400)

Predominant cellular population is admixed with scattered mature adipocytes and with stellate/ spindled mesenchymal cells. Tumoral stroma has a myxoid character. A distinctive feature of this neoplasm is the existence of a vascular network composed of curved thin-walled capillaries with a branching pattern ("chicken-wire") (Figure 5). The mitotic activity is low. The surface epithelium shows polypoid hyperplasia alternating with areas of atrophy.

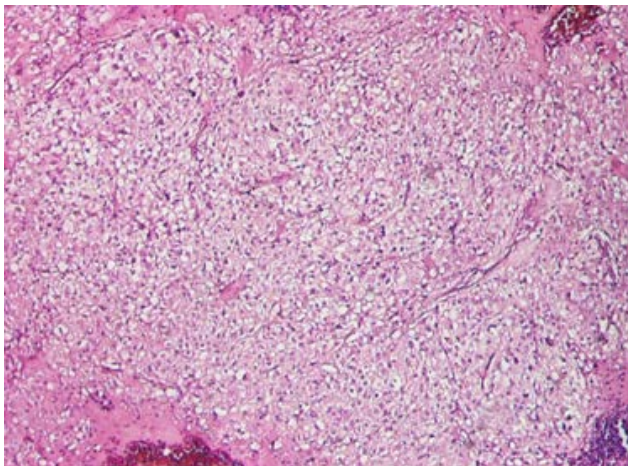

Figure 5. Chicken-wire pattern of vascularisation (H.E. X40)

The histopathological traits and the immunohistochemical profile of this tumor, characterized by positive reaction to Vimentin and S100 and negative staining to pancytokeratin, support the diagnosis of myxoid liposarcoma.

Discussions

Liposarcoma is a common soft tissue malignant neoplasm representing 15%-20% of all sarcomas and affecting almost exclusively the adults with a peak incidence in the sixth and seventh decade of life. It is commonly located in the limbs, retroperitoneal space, trunk and extremely rare in viscera [2]. The incidence of gastrointestinal liposarcoma discovered at autopsy is ranging between 0.1% and 5.8% [5]. Gastric involvement by liposarcoma is exceptional, only a few cases being recorded in international medical scientific publications. Its microscopical distinctive attribute is the presence of lipoblasts (resemble fetal adipous cells) [2].

According to WHO, the main histopathological subtypes of liposarcoma are as follows: (1) well-differentiated liposarcoma; (2) dedifferentiated liposarcoma; (3) myxoid liposarcoma; (4) round cell

liposarcoma; (5) pleomorphic liposarcoma [6].

Our case proved to be myxoid liposarcoma, a tumor consisting of primitive mesenchymal cells, lipoblasts and an abundant mucoid matrix with a particular vascular component (curved thin-walled arborizing capillaries) [7].

The rare incidence of this tumor in the stomach requires a careful microscopical examination, in order to differentiate it from other clinico-pathological entities and to avoid diagnostic errors.

Lipoblastoma is characterised by a similar histology, but it represents a tumor of infancy [7].

Round cell liposarcoma (poorly differentiated myxoid liposarcoma) is a high-grade neoplasm consisting mainly of aggregates of small round/ spindle cells with low amount of eosinophilic granular cytoplasm and large nuclei; lipoblasts are sparse; mitotic activity is intense; vascular component is not as well represented as in myxoid liposarcoma [7].

Myxofibrosarcoma is an ill-defined tumor involving mostly subcutaneous tissue of the extremities and skeletal muscle. Histopathologically, it is composed of a myxoid stroma containing cells that resemble lipoblasts or myofibroblasts with various degrees of differentiation and with a perivascular arrangement; the cytoplasmic fat vacuoles are absent; the vascular walls are thicker and the vessel shape is curvilinear; the presence of atypical mitoses, necrosis, hemorrhage and bizarre multinucleated cells is evident [7].

Myxoma is a benign tumor made of an abundant pale basophilic matrix in which rare bland spindle cells can be noticed; lipoblasts are absent; vascularity is reduced. Immunohistochemically, myxoma is positive to Vimentin, but reveals a negative/a very rare positive immunostaining to S100 [6,7].

Epithelioid gastric stromal tumor is characterized microscopically by sheets of plump epithelioid cells with eosinophilic cytoplasm that show perinuclear condensation and peripheral clearing, round nuclei; in some cases, these cells could be admixed with isolated multinucleated giant cells; stroma exhibits hyaline degeneration, liquefaction or areas with rich mucopolysaccharidic content. Immunohistochemical profile includes positive reaction to CD117, CD34 and Vimentin [8].

Diffuse type gastric carcinoma consists of a population of mucin-containing signet ring cells (versus lipid-containing signet ring cells in liposarcoma) that originate in the mucosa and infiltrate transmural as individual cells. Special histochemical stainings for mucin and lipids differentiate the two lesional entities, as well as the immunoexpression for cytokeratin in carcinoma [9].

Gastric metastasis can be excluded by the interpretation of anamnestic data (history of carcinoma) and by the immunoreactivity to cytokeratin.

After thorough analysis of all histopathological characteristics of the presented case and after differentiation from other lesional entities, the final diagnosis of myxoid liposarcoma imposed a complex therapeutic strategy. After two years of follow-up, the health status of the patient is favorable, without recurrences.

Conclusions

Myxoid liposarcoma is an extremely rare tumor in gastric location and may present common histopathological characteristics with other neoplasms of mesenchymal or epithelial origin. Correlation of clinical, imagistic, morphological and immunohistochemical data is mandatory in order to establish an accurate histopathological diagnosis and to initiate targeted therapy.

References

1. Laky, D. & Stoica, T. (1986). Gastric liposarcoma. A case report. *Pathol Res Pract*. 181(1), 112-7. PMID: 3703739 [PubMed - indexed for MEDLINE]

2. Tepetes, K., Christodoulidis, G., Spyridakis, M.E., Nakou, M., Koukoulis G. & Hatzitheofilou, K. (2007). Liposarcoma of the stomach: A rare case report. *World J Gastroenterol.* 13(30), 4154-4155, PMID:17696242 [PubMed - indexed for MEDLINE]
3. Seki, K., Hasegawa, T., Konegawa, R., Hizawa, K. & Sano, T. (1998). Primary liposarcoma of the stomach: a case report and a review of the literature. *Jpn J Clin Oncol.* 28(4), 284-8
4. Fletcher, C.D.M. (2007). *Diagnostic histopathology of tumors* (3rd edition). Churchill Livingstone
5. Yang, B., Shi, P., Li, X. & Xu, R. (2006). Well-differentiated liposarcoma of esophagus. *Chinese Medical Journal.* 119 No. 5: 438-440. PMID: 16542592 [PubMed - indexed for MEDLINE]
6. Fletcher, C.D.M., Unni K.K. & Mertens F. (2006). *Pathology and Genetics of Tumors of Soft Tissue and Bone* (3rd edition). World Health Organization
7. Weiss, S.W. & Goldblum, J.R. (2008). *Soft tissue tumors* (5th edition) (pp. 312-1080). Mosby Elsevier
8. Miettinen, M. & Lasota, J. (2006). Gastrointestinal stromal tumors: review on morphology, molecular pathology, prognosis and differential diagnosis. *Arch Pathol Lab Med*. 130(10), 1466–1478. PMID:17090188 [PubMed - indexed for MEDLINE]
9. Bosman, F.T., Carneiro, F, Hruban, R.H. & Theise, N.D. (2010). *WHO Classification of Tumours of the Digestive System* (4th edition). World Health Organization