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CASE REPORT

Posttraumatic intraosseous epidermoid inclusion cyst. A case report and short literature review

Adrian Dumitru* **, Mariana Costache* **, Bogdan Șerban*, Ioan-Mihai Japie***, Dan Anghelescu***, Constantin Condrea****, Doris Ionescu**, Cătălin Aliuș*****, Sebastian Grădinaru** *****

*Department of Pathology, University Emergency Hospital Bucharest, Romania
**"Carol Davila" University of Medicine and Pharmacy, Bucharest, Romania
***Department of Orthopaedics and Traumatology, University Emergency Hospital Bucharest
****Department of Plastic Surgery, University Emergency Hospital Bucharest, Romania
*****4th Surgery Department, University Emergency Hospital Bucharest, Romania

Correspondence to: Adrian Dumitru,
Department of Pathology, University Emergency Hospital Bucharest, Romania,
169 Splaiul Independenței Street, code 050098, Bucharest, Romania,
Phone: +40213 180 523, E-mail: dr.adriandumitru@yahoo.com

Abstract

Epidermoid bone inclusion cysts are pathological structures bordered by a layer of stratified squamous epithelium containing keratin debris. They are also called epidermal bone cysts. Intraosseous epidermoid cysts are rare entities; these lesions may occur as a result of trauma (puncture wound or pressure erosion).

They appear to be well defined, translucent lytic lesions with sclerotic margins on imaging studies. The most frequent sites affected are: the distal phalanx and the calvaria - intradiploic, but a few cases of larger bone involvement have been reported. The imagistic differential diagnosis of this lesion includes intraosseous extension of a glomus tumor, enchondroma, osteoid osteoma, acral metastases, some primary vascular bone tumors, giant cell reparative granuloma, subungual nodular melanoma or other lytic non-neoplastic lesions such as osteomyelitis. We present the case of a 70-year-old man experiencing anxiety and pain due to distal phalanx expansion of the IV digit, following hand trauma. An X-ray was performed, showing a radiotransparent lesion located in the distal phalanx. An intramedullary inclusion cyst must be taken into account as a clinical differential diagnosis. Due to the extensive destruction of the distal phalanx, amputation was necessary. Gross examination of the surgical specimen revealed a cystic lesion that contained whitish, creamy material with extensive necrotic areas. Microscopic examination revealed a ruptured intraosseous epidermoid inclusion cyst, which was successfully excised.

Keywords: bone, cyst, epidermoid, posttraumatic, histopathology

Introduction

Intraosseous epidermoid inclusion cyst, also known as epidermoid bone cyst, is usually an unilocular cyst located intramedullary, containing keratin, and being covered by a squamous epithelium. Adult males are affected most frequently [**1,6**]. Most

intraosseous epidermoid inclusion cysts are acral lesions often found in manual workers, usually related to previous trauma. These lesions commonly occur in males in the third to sixth decades [**1,2**]. Skull lesions are supposed to be congenital lesions and are more frequently diagnosed in the first decade of life [**3**]. Those lesions are considered to arise from ectodermal inclusion during neural tube closure and subsequently remain within the cranial bones. The vast majority of these lesions are located in the distal phalanges of the upper limbs, with some notable exceptions such as the jaws, the skull and the sacral bone [**1,4,5,9**].

Radiologically, epidermoid bone cysts are well-defined, round osteolytic lesions [**7**]. Histopathologically, those lesions consist of an inner lining of squamous epithelium with a granular layer covered by sheets of laminated keratin that occupy the whole cavity [**8**]. Therapeutic options are either curettage or excision depending on the clinical presentation. However, some lesions are difficult to diagnose because they can mimic a number of entities, so a rigorous clinical history and a differential diagnosis are always mandatory. Due to the rarity of the disease, it is difficult to establish the correct diagnosis before surgery [**9**].

Case Report

A 70-year-old man suffered a bicycle accident 7 years prior to this presentation. He injured his back, his right hand and shoulder. While his back and shoulder did not require surgical treatment, his right hand was badly injured. The patient lost the middle finger distal phalanx and his fourth finger showed only small abrasions and slight crushing of the nail bed. At that time, no fractures or other bone lesions were identified. After a few weeks, the patient was able to resume work and the function of his right hand was not affected. The distal phalanx of the IV digit had gradually enlarged over the next 7 years, causing intermittent pain as it could not fit into his working glove anymore. Eventually, the pain worsened, especially during the night, and the patient went to the doctor. A hand X-ray was performed, revealing a round radiolucent lesion with a "ground glass" appearance in the distal phalanx. Also, the film showed an extensive destruction of most of the distal phalanx. Apart from the accident that occurred 7 years before, the patient had no medical history for other pathology. The clinical examination revealed a tumefaction of his fourth finger and enlargement and tenderness of the nail (**Fig. 1**).

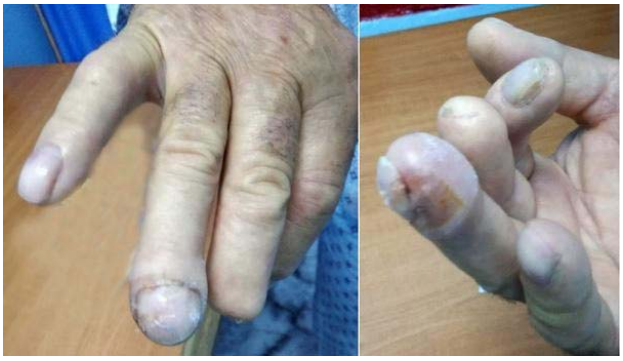

Fig. 1 The distal phalanx of the IV finger is notably enlarged and tender. The skin shows periungual soft tissue oedema and hyperemia resembling an infection. What should be noted is that the distal phalanx of the IIIrd finger is missing due to previous trauma

Clinically, a giant cell tumor or an osteoid osteoma aggravated by his previous injury were suspected. Due to extensive injuries to the distal phalanx and given that curettage of the lesion was no longer an option, amputation of the affected area was recommended. Postoperative evolution was favorable with the preservation of the normal function of the hand.

Material and methods

The specimen obtained after successful surgical excision was sent to the Pathology

Department for histopathological assessment. The gross examination revealed a well-defined unilocular cystic cavity with a diameter of 1,6 cm. The inner wall was about 2 mm thick. The cystic cavity contained a yellowish amorphous substance. The neighboring bone tissue showed small hemorrhagic areas. After a slight decalcification with a chelating agent (ethylenediaminetetraacetic acid/ EDTA), the specimen was processed according to the usual histopathological method. Decalcification was necessary to obtain soft sections of the bone using a microtome. Fixation was carried out using 10% buffered formalin. The samples were processed by paraffin inclusion and conventional Hematoxylin–Eosin (HE) staining.

Results

Histopathological examination revealed a cystic lesion filled with keratin debris (**Fig. 2**). The cystic wall was made up of a thin keratinizing stratified squamous epithelium having a distinct granulosa layer (**Fig. 3**).

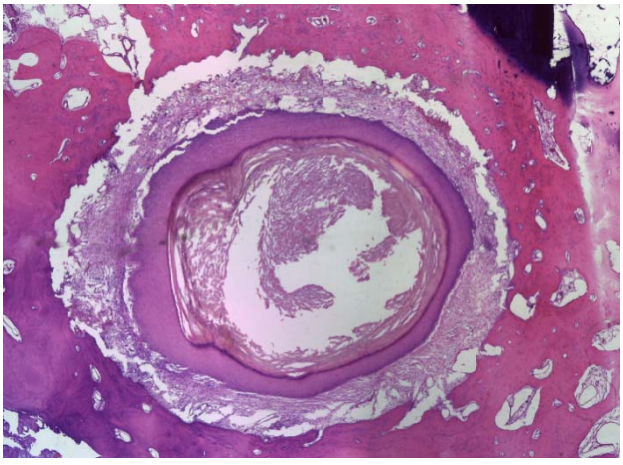

Fig. 2 Intraosseous unilocular cystic lesion composed of a thickened wall of stratified squamous epithelium. The cystic cavity is partially filled with keratin debris. The bone tissue at the periphery is sclerotic. Hematoxylin-eosin staining, original magnification 40x. Stitch image using Leica Application Suite software

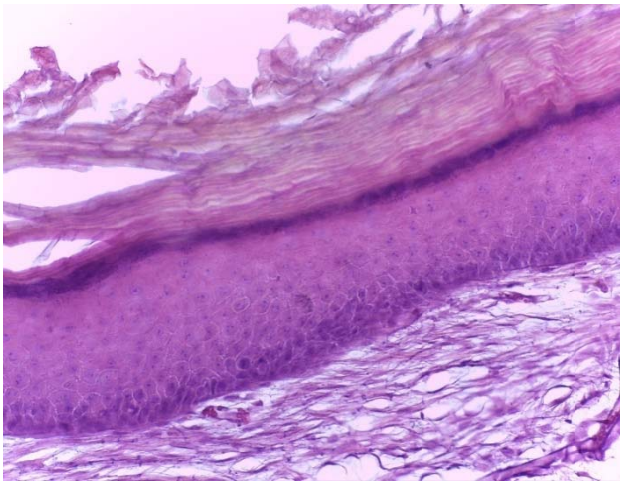

Fig. 3 Detailed histopathological section of the cystic wall showing keratinized stratified squamous epithelium with a separate granular layer and abundant keratin debris. Hematoxylin-eosin staining, original magnification 100x

The squamous epithelium had few cellular layers and was separated from the cortical bone tissue by a layer of connective tissue displaying a moderate inflammatory response. Also, we observed some areas of osteoclastic endosteal reabsorption in the vicinity of the cystic cavity. A portion of the cystic lesion was ruptured with an extensive foreign body inflammatory reaction nearby. The inflammatory reaction was composed of multinucleated macrophages around devitalized keratin lamellae and cholesterol crystals. The cystic lesion did not make any contact with the nearby epidermis. Considering the suggestive histopathological aspects, the diagnosis of posttraumatic intraosseous epidermoid inclusion cyst was made.

Discussions

Epidermoid inclusion cysts are slow-growing, benign, painless masses commonly seen in skin or subcutaneous soft tissues. Only a handful of cases of intraosseous inclusion epidermoid cysts are reported in literature.

They rarely appear in the bones, usually under the form of pseudotumors or small lytic lesions, either in the skull of young patients or in the phalanges of older patients. Also, there are some reports of epidermoid bone cysts of temporomandibular joint, mandible, maxilla, vertebrae, tibia, sacrum and femur [**1,3,5,8,9**]. Apart from the location in the distal phalanges of the fingers, posttraumatic epidermoid cysts have been reported in only 4 cases involving the great toes [**10**]. Although most lesions are painless, some patients experience tenderness and pain and seek medical attention particularly in the case of large, broken or recently traumatized cysts. Male patients are more predisposed to the disease, especially those who perform manual work and are more prone to injury [**1,4,6,9,11**]. The distal phalanges of the fingers, particularly the left middle fingers, are more commonly affected.

Most intraosseous epidermoid cysts are solitary lesions, but there are rare cases of patients who presented with cysts affecting multiple fingers of the same hand. It is believed that the appearance of these lesions is secondary to a traumatic event, being caused by the intrusion of epidermal tissue into the bone [**1-5,9,11**]. Lesions of this kind are relatively easy to diagnose if the patient's personal history is taken into account. The first step in establishing a correct diagnosis is a careful clinical examination and radiological studies. The classical radiologic presentation of the intraosseous epidermoid cyst is as a well-circumscribed, expanding radiolucent bone lesion, often associated soft tissue oedema [**7,9,10,12**]. The cortical bone can suffer remodeling and expansion due to these cysts. MRI imaging of epidermoid cysts reveals a well-defined mass with high T2 signal. Contrast enhancing T1W imaging reveals a thin margin intensification [**12,13**]. MRI is useful especially in differentiating other injuries as well as in assessing the invasive character in the surrounding tissue, in spite of its decreased ability in defining bone alterations [**14**].

In case of lesions with unusual presentation or with atypical radiological aspects, other pathological entities can be taken into account: chronic bone and soft tissue infections, benign as well as malignant tumors, including aneurysmal bone cyst, osteoid osteoma, simple bone cyst, chondroma, giant cell tumor of the bone, some reparative lesions and, in extremely rare cases, even metastatic lesions [**8,10**]. The benign cartilaginous tumor, such as enchondroma, is the most commonly found phalangeal lesion, presenting as a spotty calcification in a radiolucent area. In our case, the lesion was more clinically similar to an osteoid osteoma, another entity that should be taken into account as a differential diagnosis. Osteoid osteoma is an X-ray easily identifiable lesion and is associated with pain and perifocal reactive sclerosis. A slow growing, painless lesion with surrounding sclerosis frequently allows an easy radiologic diagnosis of these cysts, especially in patients with a history of hand trauma. Some other entities such as aneurysmal giant cell tumor of the bone, bone cyst and intraosseous extension of a soft tissue glomus tumor may exhibit radiological similarities with intraosseous epidermoid cyst [**4,8,10**].

Regarding the differential diagnosis with bone metastases, although cases of this type have been reported from occult malignancies (such as lung cancer), it is very unusual for such lesions to present as the first clinical symptom [**15**]. In these particular circumstances, manifestations of the primary neoplasia are usually present.

Many pseudotumors can be described as vaguely defined lytic lesions on imagistic investigation (osteomyelitis and gout) but such cases are usually accompanied by suggestive clinical features [**4,10**].

Nevertheless, the correct diagnosis is established by histopathological examination.

In some cases, frozen section diagnosis can be performed and it is a very valuable tool because once the benignity of the lesion is certified, amputation of the affected segment can be avoided [**10**]. Fine needle aspiration can be another useful tool for the diagnosis of intraosseous epidermoid cyst of the terminal phalanx [**16,17**]. In such ideal cases, curettage of the cyst can be performed followed by bone grafting. This is preferable as it can avoid other invasive treatment. However, amputation is inevitable when the lesion is too large, ruptured, infected or the architecture of the finger is compromised [**10**]. Painful cysts should be treated early for both cosmetic and functional considerations.

After surgical removal, all samples should be sent to the histopathological department in order to confirm the correct diagnosis.

Conclusion

In summary, intraosseous epidermoid inclusion cysts are rare clinical entities that have a history of previous trauma. Radiolucent osseous neoplasms, benign or malignant should be included in the differential diagnosis. This case report emphasized, for both clinicians and pathologists, the importance to have a high index of suspicion regarding radiolucent bone lesions of the distal phalanx, especially in patients with previous trauma and the diagnosis of intraosseous epidermoid cyst should always be considered.

Conflict of Interest statements

Authors state no conflict of interest.

Informed Consent and Human and Animal Rights statements

Informed consent has been obtained from all individuals included in this study.

Authorization for the use of human subjects

Ethical approval: The research related to human use complies with all the relevant national regulations, institutional policies, is in accordance with the tenets of the Helsinki Declaration, and has been approved by the authors' institutional review board or equivalent commitee.

Ethical standards

We undersign and certificate that the procedures and the experiments we have done respect the ethical standards of the Helsinki Declaration of 1975, as revised in 2000, as well as the national law.

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