Rare disease

A suspected malignancy in osteolytic bone tumour of the thumb

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A 75-year-old male patient was referred to our institution owing to a painful and gradually developing lesion of the thumb with suspicious malignancy. The patient was suffering from a swollen, red, tender left thumb for 3 months. An old scar at the finger pulp could be traced from an old minor trauma. The x-ray revealed an osteolytic lesion in the terminal phalanx of the non-dominant hand that raised concerns of malignancy. Additional investigations such as ultrasound, CT-scan and MRI have been performed to get better insight to the lesion. After performing a biopsy, no malignant cells were found. Owing to the local destroying effect of the lesion and the clinical signs of the patient, the lesion was excised in total. The histopathological evaluation confirmed the tumour as a rare intraosseous epidermoid cyst. A bone graft after resection was not needed. The postoperative follow-up of the patient was uneventful.

BACKGROUND

Osteolytic bone tumours of the hand can be seen frequently as incidental findings in x-rays even in asymptomatic patients. The majority of the lesions are benign but, in rare cases, the bone tumour can be malignant. Clinical signs are unspecific and vary from redness, swelling and tenderness to complete asymptomatic patients. Sometimes, a fracture due to the thinning of the bone density can initiate symptoms. Further evaluations include x-rays and, if necessary, ultrasound, CT scans or MRI, in order to facilitate therapeutic decisions. Needle biopsy can provide additional preoperative information.

CASE PRESENTATION

A 75-year-old man with 3 months gradually growing swelling of the terminal phalanx of the thumb was referred to our institution. The patients' history was unremarkable except an old scar at the pulp of the left thumb resulting from a crushing minor trauma to the top of the finger and laceration of the pulp more than 5 years ago. After suturing of the laceration, the injury had healed spontaneously without evidence of infection. There was no previous malignant or other systemic disease. Physical examination at the time of admission was negative except for the distal segment of the fingertip. The x-ray showed an intraosseous lesion of the terminal phalanx raising concerns of malignancy (figures 1 and 2). After supplementary evaluation with ultrasound, CT scan and MRI (figures 3,4,5,6,7), a biopsy was performed that revealed a squamous epithelium. No malignant cells were found. After curettage (figures 8 and 9) and histological examination showing the cyst wall to be stratified squamous epithelium with keratinised fragments with no evidence of neoplasia, the diagnosis was confirmed as an epidermoid cyst (figures 10 and 11).

Figure 1 x-Ray at admission with osteolytic lesion of the terminal phalanx (anterior-posterior and lateral view).

Figure 2 x-Ray at admission with osteolytic lesion of the terminal phalanx (anterior-posterior and lateral view).

INVESTIGATIONS

In the radiographs, a radiolucency of the terminal phalanx of the thumb was described. The cortical bone was eroded in the palmar aspect of the bone without trabecular bone structure. A periosteal reaction was not evident. A CT

Figure 3 Ultrasound evaluation of the tumour.

Figure 4 Preoperative CT scan of the thumb to get better insight to the amount of bone loss.

scan was performed to assess the dimension of the bone defect. Calcifications inside the tumour were not present while it revealed a plain encapsulation of the lesion. Furthermore MRI showed 'onion ring'-like structures in

Figure 5 Preoperative CT scan of the thumb to get better insight to the amount of bone loss.

Figure 6 Non-enhancing onion ring- like structure of the thumb in MRI.

the lesion. After excision and curettage, histopathological examination confirmed the preoperative suspicion of an epidermoid bone cyst.

DIFFERENTIAL DIAGNOSIS

Bone cysts of the hand can be challenging both clinically and radiologically. The differential diagnosis includes benign and malign bone tumours. In x-rays, a distinction has to be drawn between well-defined and poor-defined lesions. The most frequently well-defined osteolytic lesion is the enchondroma predominantly located in the metacarpal bone or proximal phalanx. The tumour may have

Figure 7 Non-enhancing onion ring-like structure of the thumb in MRI.

Figure 8 Intraoperative picture of the palmar aspect of the left thumb after resection of the lesion with cavity and remaining parts of the capsule.

calcifications but does not show periosteal alterations. The lack of periosteal reactions should rule out an inflammatory lesion such as an osteomyelitis. Another radiologically well-defined entity is the giant cell tumour. It occurs most frequently in the terminal phalanx and grows locally aggressive with destruction of the bone, although the lesion itself is benign. Another well-defined lesion with sclerotic margins is the glomus tumour that occurs subungually. The tumour is painful, raises tenderness and cold intolerance. Poor-defined lesions can be metastatic or primary tumours. Metastases are rarely found and mostly deposits of breast or bronchus primary. The occurrence of Ewing sarcoma has also been described in the terminal phalanx.¹ Aneurysmal bone cyst has to be considered in extensive osteolytic changes. Clinically, the epidermoid cyst can mimic psoriasis, malignancy or monoarticular arthritis.²⁻⁴

TREATMENT

An open biopsy was performed in block anaesthesia with a palmar approach along the scar in order to remove any scar tissue. Dissection was carried down the terminal

Figure 9 Intraoperative picture of the palmar aspect of the left thumb after resection of the lesion with cavity and remaining parts of the capsule.

Figure 10 (A) A few keratin lamellae from the cyst wall. (B) Lymphohistiocytic inflammatory infiltrate. (C) Adjacent bone tissue (H&E stain; magnification $\times 200$).

phalanx. The remaining cortical bone was removed. A yellow-brownish 'cheese-like' 1.5 cm in diameter holding mass with a thin capsule layer was peeled off in total that left a cavity. The specimen was sent to histological evaluation. A bone graft was not implanted (figures 12 and 13).

OUTCOME AND FOLLOW-UP

The postoperative healing of the wound was uneventful and the patient regained full function 6 weeks after operation. The histopathological result provided diagnose of

Figure 11 Stratified squamous epithelial capsule with keratin lamellae (H&E stain, magnification $\times 100$).

an intraosseous epidermoid cyst with a stratified squamous epithelial capsule and centrally deposited keratin. As we have excised the whole lesion, a recurrence is not expected.

DISCUSSION

The epidermoid cyst is a non-neoplastic lesion characterised histologically by a membrane consisting of squamous epithelium, covered by laminated masses of keratin that may occupy part of the cavity.⁵

The origin of intraosseous epidermoid cyst remains unclear. Epidermoid cysts arise most commonly in the skull but occur also in the tibia, ulna and femur. They have been described in 1923 by Sonntag.^{6 7} It can either develop after an injury with inoculation of intraosseous squamous cells⁸ as it was the case in our patient. It can also be due to a failed embryogenetic differentiation.⁹ It is most common in male adults and in the terminal phalanx of the finger.¹⁰ The incidence is unclear but 4 intraosseous of 101 epidermoid cysts of the hand indicate a very low incidence.¹¹

Clinically, the lesion can have variable degrees of redness, tenderness and swelling. Even complete asymptomatic patients can be seen. Pathological fractures resulting from a weakening of the bone strength can initiate symptoms.

Epidermoid cysts have to be removed in total. The results after complete resection are excellent. Amputation should be reserved for almost complete destruction of the

Figure 12 Postoperative x-ray of the thumb with resected bone tumour of the terminal phalanx with lack of palmar cortical bone due to the destructive character of the epitheloid cyst (anterior-posterior and lateral view).

Figure 13 Postoperative x-ray of the thumb with resected bone tumour of the terminal phalanx with lack of palmar cortical bone due to the destructive character of the epitheloid cyst (anterior-posterior and lateral view).

phalanx and every effort should be made to restore a structurally stable, tactilely competent distal phalanx.⁶

Bone tumours can be seen frequently even in asymptomatic patients. Further evaluation is needed, especially in poor-defined, expanding osteolytic lesions. Ultrasound and CT scan evaluation can be advantageous for a preoperative evaluation. MRI is recommended in any suspicious case and is the recommended approach.¹² The epidermoid cyst is not contrast-enhanced in MRI.¹³ A needle biopsy in existing suspicious malignancy is supportive for planning the degree of surgical intervention. In malign lesions and local destructing tumours, a resection in total is of utmost importance to prevent recurrence.

Learning points

- Intraosseous epidermoid cysts can present with variable symptoms, predominantly occur in male patients and develop mostly, but not exclusively, after trauma.
- Bony lesions in x-rays can be seen by coincidence. Poor-defined lesions and cases of suspected malignancy have to be evaluated.
- Radiological evaluation can be based on x-ray, ultrasound, CT-scan or MRI. A fine needle biopsy is valuable in unclear cases to provide a preoperative histological diagnose.
- The radiological characteristics of intraosseous epidermoid cysts are radiolucency without internal calcifications and the lack of periosteal signs. There is no enhancement in the MRI. The prognosis after complete resection is excellent.

Competing interests None

Patient consent Obtained.

REFERENCES

1. **Strege DW**, Hanel DP, Vogler C, *et al.* Ewing sarcoma in a phalanx of an infant's finger. A case report. *J Bone Joint Surg Am* 1989;**71**:1262–5.
2. **Bhagwandas K**, Nicolaou N, Roberts DL. Intraosseous epidermoid cyst mimicking psoriasis. *Br J Dermatol* 2001;**145**:366–8.
3. **Pagnano MW**, Athanasian EA, Bishop AT, *et al.* Intraosseous epidermoid cyst in a metacarpal mimicking malignancy. *Orthopedics* 1997;**20**:719–21.
4. **Radke S**, Walther M, Ettl V, *et al.* Epidermoid cyst mimicking monoarticular arthritis of the great toe. *Rheumatol Int* 2004;**24**:117–19.
5. **Schajowicz F**, Sissons HA, Sobin LH. The World Health Organization's histologic classification of bone tumors. A commentary on the second edition. *Cancer* 1995;**75**:1208–14.
6. **Hinrichs RA**. Epidermoid cyst of the terminal phalanx of the hand. Case report and brief review. *JAMA* 1965;**194**:1253–4.
7. **Sonntag K**. [Traumatische Epithelzyste im Knochenende an einem Fingerstumpf als Unfallfolge.] *Munchen Med Wschr* 1923;**70**:1055–6.
8. **Lucas GL**. Epidermoid inclusion cysts of the hand. *J South Orthop Assoc* 1999;**8**:188–92.
9. **Niccoli C**, Mambelli V. (Intra-osseous epithelial cyst of the phalanx). *Arch Putti Chir Organi Mov* 1978;**29**:395–404.
10. **Hamad AT**, Kumar A, Anand, *et al.* Intraosseous epidermoid cyst of the finger phalanx: a case report. *J Orthop Surg (Hong Kong)* 2006;**14**:340–2.
11. **Lincoski CJ**, Bush DC, Millon SJ. Epidermoid cysts in the hand. *J Hand Surg Eur Vol* 2009;**34**:792–6.
12. **Simon K**, Leithner A, Bodo K, *et al.* Intraosseous epidermoid cysts of the hand skeleton: a series of eight patients. *J Hand Surg Eur Vol* 2011;**36**:376–8.
13. **Nakajo M**, Ohkubo K, Nandate T, *et al.* Intraosseous epidermal cyst of the distal phalanx of the thumb: radiographic and magnetic resonance imaging findings. *Radiat Med* 2005;**23**:128–32.

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