Rare disease

Adamantinoma of the tibia and fibula with pulmonary metastasis: an unusual presentation

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Summary

Adamantinoma is a rare tumour of long bones, representing less than 1% of them. Adamantinoma commonly occurs in the tibia. It is locally aggressive and recurrences are uncommon after resection. Metastases have been reported in less than 10% of cases. The most common radiographic appearance is multiple sharply demarcated radiolucent lesions surrounded by areas of dense sclerotic bone. The authors report a patient who developed pulmonary metastasis 1 year after complete resection of primary neoplasm.

BACKGROUND

Adamantinoma is one of the most low-grade neoplasm that affects long bones and accounts for less than 1% of all primary bone tumours.^{1–3} Adamantinoma was described initially by Fisher in 1913.^{3–4} Adamantinoma occurs most commonly between 10 and 50 years with slight male preponderance. It usually arises in the centre of long bones, but may involve the whole width of the bone. Adamantinomas are slow growing tumours with a limited propensity for metastasis and local recurrence after wide resection.⁵

CASE PRESENTATION

A 50-year-old woman presented with a history of pain and swelling of mid part of the left leg since 6 months without history of trauma. Clinical examination showed an irregular swelling with a lobulated surface of variable consistency and collateral venous circulation. The skin over the swelling was stretched and shiny. Mobility of the knee was painful and limited.

INVESTIGATIONS

X-rays exploration showed an expansive demarcated radiolucent lesion surrounded by areas of dense sclerotic bone involving the mid part of the tibia with extension to the soft tissues and periosteal reaction on the fibula diaphysis (figure 1). On MRI, the lesion showed a hypo signal on T1-weighted sequences and a heterogeneous hyper signal on T2. Injection of gadolinium showed heterogeneous contrast enhancement of the tumour. MRI showed the extra osseous involvement with large extension to the soft tissues and to the fibula (figure 2). A complementary angio-CT demonstrated popliteal artery invasion (figure 3). Skeletal scintigraphy showed increased uptake of the radionuclide on only the tibia and the fibula. Moreover, there was no evidence of metastases on initial thoracic scan.

DIFFERENTIAL DIAGNOSIS

Radiological differential diagnosis arises mainly with fibrous dysplasia and osteofibrous dysplasia.

Figure 1 Lateral (A) and antero posterior (B) radiographies: sclerosis involving the tibia metadiaphysis with multiple lytic areas with extension to the soft tissues and fibular discontinuous periosteal reaction (arrowheads).

Figure 2 Sagittal T2 WI MR. (A) Expansive lesion of the tibia metadiaphysis breaking through cortex with extension to the soft tissue (A, B, C, D) and fibula (B, C, D). This mass has an increased T2 signal (A, C), low signal intensity on T1 (B) and heterogeneous intense enhancement after gadolinium injection (D).

TREATMENT

A surgical biopsy was then performed. Histology (figure 4), showed islands of epithelial cells in a fibrous stroma with chondroid differentiation, tumorous proliferation and sheets of squamous epithelium with keratinisation (figure 5). Immunohistochemistry showed positivity to cytokeratin and vimentin. These findings were consistent with a diagnosis of adamantinoma.

Above knee amputation was performed as there was knee involvement and wide involvement of muscles and all vascular axes of the leg. Histology confirmed the diagnosis of adamantinoma, and the integrity of the limits of resection. Clinical examination and radiographic investigations were normal with no evidence of recurrence.

OUTCOME AND FOLLOW-UP

The patient returned after 1 year with dyspnoea and haemoptysis. Thoracic scan showed pulmonary metastases (figure 6). Clinical examination and radiographic investigations showed no evidence of local recurrence.

Unresectable pulmonary metastases were treated with chemotherapy including etoposide and cisplatin. However, treatment had to be discontinued due to poor tolerance. The patient died 3 months after the discovery of metastases.

Figure 3 Angio CT. 3 D reconstruction: vascular extension to the popliteal artery (star shaped).

Figure 4 Histology of the tibial diaphysis (H&E x100): chondroid differentiation within tumorous proliferation.

DISCUSSION

Adamantinoma is a locally aggressive neoplasm. The incidence of recurrence is approximately 30% and those of metastases ranges between 10% and 20%.¹⁻³ Pain is the most common clinical manifestation and local swelling is the second clinical sign of appeal.⁴⁻⁶

Histopathologically, there are two forms of adamantinoma.^{7 8} The first one is the classic adamantinoma characterised by an association of both epithelial and osteofibrous component. Its low malignant potential is known and occurs usually in patients older than 20 years. The second form is the differentiated adamantinoma also called juvenile or osteofibrous dysplasia OFD-like. It occurs almost exclusively in patients younger than 20 years. Histologically, OFD-like pattern lacks conspicuous nests and masses of epithelial cells with scarred positivity of epithelial elements for cytokeratin. This form is relatively benign.

On radiographs, it is typically an osteolytic lesion, mono or multi-loculated, central or eccentric well-circumscribed with septa and a peripheral condensation.^{3 4 9} Adamantinoma is usually located in the anterior

Figure 5 Histology of the tibial diaphysis (H&E x400): sheets of squamous epithelium with keratinisation.

Figure 6 Chest CT: pulmonary metastases.

tibial diaphysis or metaphysis. The lesion is more often intracortical and can extend longitudinally but it can also destroy the cortex and invade the marrow cavity.^{4 9 10} The CT specifies better the characters of the lesion and can detect another tumour site invisible on the radiographs. Radiological differential diagnosis arises mainly with fibrous dysplasia and osteofibrous dysplasia.¹⁰

MRI has the interest to appreciate the major extension in height, to find a small lesion on the remote or on fibula, as well as a soft tissues extension.^{9 11}

In T1 sequence, tumour has a signal similar to that of muscles and in T2, a hyper signal often heterogeneous. In case of cystic component, a liquid-like hypersignal is observed on T2 sequences. Injection of gadolinium shows homogeneous contrast enhancement of the tumour and peripheral enhancement in the cystic forms.⁴

The spontaneous evolution is slow, but adamantinoma can also have a progressive local extension with soft tissues invasion. However, this situation remains rare.

Adamantinoma is highly radio resistant, and chemotherapy has not been shown to be effective. Surgical treatment includes en bloc resection with wide margins and limb

reconstruction. Amputation is rarely indicated when limb reconstruction after surgery is not possible.^{2 3 5 12}

After radical treatment, there is a high percentage of healing. The main factor of recurrence is incomplete resection (recurrence rate is almost of 30% with an interval of 85 months).^{2-5 7 12}

Our case was unusual because the patient developed rapid growing lesion in both tibia and fibula. Moreover, lung metastases were noted 1 year after a wide en bloc resection of the tumour with surrounding normal tissue. Some authors reported local recurrence and lung metastases after complete resection and claim that even a wide tumorous resection can give good results of healing, long-term follow-up is mandatory.^{3 5 12 13}

Learning points

- The spontaneous evolution of adamantinoma is slow, but it can also have a progressive local extension with soft tissues invasion.
- The unusual radiographic appearance of adamantinoma in our case with rapid growing and the high potential to metastasise has not been reported previously in the literature and should be known.
- Even though a wide tumorous resection usually gives good results of healing, long-term follow-up is mandatory.

Competing interests None.

Patient consent Obtained.

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