Rare disease

Ossifying fibromyxoid tumour: a rare soft tissue tumour of intermediate malignancy

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Summary

Ossifying fibromyxoid tumours (OFTs) are uncommon soft tissue tumours. A 73-year-old male presented with a painless exophytic mass located in the left scapular area which was identified 1 year earlier. An incisional biopsy was performed and microscopic features were compatible with OFT. Radiologic evaluation revealed multiple pulmonary metastasis. Although many cases pursue an indolent biologic behaviour, local recurrence and metastasis have been reported and long-term follow-up should be considered.

BACKGROUND

The ossifying fibromyxoid tumour (OFT) of soft tissue, is a rare tumour of uncertain differentiation which most commonly arises in the extremities. Close to 150 cases have been reported in the literature, mostly in the form of case reports or small series. Although the original description emphasised the bland morphologic appearance and benign clinical behaviour, several subsequent reports described tumours with typical features that unexpectedly metastasized, or tumours which had atypical or overtly malignant histologic features, some of which behaved aggressively.^{1–4} The presented case had typical morphology but had multiple pulmonary metastasis at presentation.

CASE PRESENTATION

A 73-year-old farmer with left scapular mass was admitted to the surgery ward of Shahid Sadoughi General Hospital in Yazd (the centre of Yazd province in the central part of Iran). The scapular mass had been noticed 1 year ago and had growth during time (figure 1). There was no medical or surgical problem except mild exertional dyspnoea. On general appearance, the patient appeared normal and alert and had normal vital sign. There was no mass, pigmentation or ulceration on the other parts of the body. On local examination, there was a large left-sided scapular mass with extension to the supraclavicular area, measure about 15 cm. The mass was slightly movable, well-defined and multilobular with elastic consistency. The mentioned mass showed no sign of inflammation. There was not any other abnormality in systemic examination. The laboratory tests were within normal limits. Radiologic evaluation revealed a large mass that encased the scapula without significant bone erosion with scattered calcification and multiple lung nodules. These nodules were 2–3 cm in diameter and were scattered throughout both lungs. Incisional biopsy from scapular lesion was performed, microscopically the lesion consisted of bland small to medium sized cell in a fibromyxoid background and a thick collagenous capsule containing an incomplete rim

of lamellar bone. There was no significant mitotic activity or necrosis (figures 2 and 3). The tumour was positive to s100, NSE, GFAP and vimentin. The definitive diagnosis was OFT. Owing to, the patient was relatively asymptomatic and there was no chemotherapeutic planning, these nodules were not biopsied.

DIFFERENTIAL DIAGNOSIS

The differential diagnosis includes benign and malignant epithelioid nerve sheath tumour, chondroid syringoma, myxoid chondrosarcoma and epithelioid smooth muscle tumours.

TREATMENT

Generally, it has been suggested that tumour resection and regular clinical follow-up are considered to be the appropriate management of the tumour at present.^{5–8} There is no consensus about adjuvant therapies. Some believe that adjuvant therapy does not seem to be indicated,¹ while others have suggested that treatment should consist of wide surgical excision of primary tumour and radiotherapy. In addition, further clinicopathological investigation of the OFT has to be a prerequisite to obtain an appropriate treatment of the tumour. Early follow-up to discover early local recurrences or metastasis is mandatory. Nevertheless, our patient did not receive any treatment because his tumour was bulky, he had pulmonary metastasis and he was relatively asymptomatic.

OUTCOME AND FOLLOW-UP

The patient is in good condition after 5 months follow-up and we do not have any additional imaging.

DISCUSSION

OFT almost exclusively affects adults (mean age near 50 years) with rare examples in children.^{9 10} Men are affected more commonly than women. Most patients present with a small, painless, well-defined, often lobulated

Figure 1 Gross male patient with OFT.

Figure 2 Fibrous zone in an OFT with cords of tumour cells (H&E $\times 40$).

subcutaneous mass which involves the extremities. Grossly OFT is usually well circumscribed, covered by a thick fibrous pseudocapsule. Most measure 3–5 cm, but occasional lesions are 15 cm or larger.¹¹ Microscopically, the majority are located in the subcutaneous tissue but some are attached to tendons, fascia or involve the underlying skeletal muscle. The tumour is composed of uniform round ovoid to spindle-shaped cell arranged in nests or cords and deposited in the variably myxoid and collagenous stroma. In approximately 80% of cases, there is an

incomplete shell of lamellar bone found at the periphery of the nodules, but up to 20% of cases are non-ossifying.^{12 13} Immunohistochemically, the cells are positive for vimentin and express S100 protein in about 70% of cases the cells may also express leu7, neuron specific enolase and glial fibrillar acidic protein. The histogenesis of this tumour is controversial but Min *et al*¹⁴ suggested the cell have the ultrastructural features of myoepithelial cells. The differential diagnosis includes benign and malignant epithelioid nerve sheath tumours (epithelioid neurofibroma,

Figure 3 Trichrome staining in OFT (×40).

epithelioid schwannoma, epithelioid malignant peripheral nerve sheath tumour), chondroid syringoma (cutaneous mixed tumour), myxoid chondrosarcoma and epithelioid smooth muscle tumours.

Behaviour of the OFT varies. The vast majority of these tumours are histologically benign. Not unexpectedly most of these pursue a benign clinical course. However, it has been noted that on rare occasion even histologically typical tumours may locally recur or metastasize. This is true for our case. Yoshida and colleagues² described a case lacking malignant features but which locally recurred, metastasised and ultimately killed the patient. Some of these tumours have atypical histologic features including high nuclear grade, high cellularity and mitotic activity of >2 MF/50 HPF.¹⁴ It is suggested that tumours with these atypical features should be regarded as sarcomas with significant potential for metastasis. But since cases that deviated from typical OFT had a metastatic rate (6%) that was similar to typical tumours (4%), Folpe and Weiss¹³ opted to avoid the term atypical OFT and considered those cases within the general category of OFT.

Learning points

- ▶ OFT is considered to be of mesenchymal origin. Most of these tumours pursue a benign clinical course but on rare occasion even histologically typical tumours may locally recur or metastasize, primary in the lung even several years after surgical removal. Because of rarity, the recognition of this entity is important.
- ▶ OFT is a soft tissue tumour of intermediate malignancy of uncertain type.
- ▶ There is no consensus about appropriate treatment at present.

Competing interests None.

Patient consent Obtained.

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