Case report

Lung metastases of diffuse giant cell tumour of the fibular tendon sheath at the ankle: A case report


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A B S T R A C T

Diffuse giant cell tumours of the tendon sheaths are described in the literature as locally aggressive soft-tissue tumours. We report the case of a 56-year-old male with a history of multiple surgical procedures for a giant cell tumour of the fibular tendon sheath at the right ankle. The multiple recurrences prompted monitoring by positron-emission tomography, which showed lung tumours. Biopsies confirmed that the tumours were metastases from the giant cell tumour of the tendon sheath. In patients with recurrent and/or diffuse giant cell tumour, positron-emission tomography is an effective monitoring tool.

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1. Introduction

Diffuse giant cell tumour (D-GCT) of the synovial tendon sheath, described by Ginzler et al. [1] as extra-articular pigmented villonodular tenosynovitis, is characterised in the literature as a locally aggressive soft-tissue tumour. D-GCT is a rare condition that accounts for only 1.6% of all soft-tissue tumours [2]. Although any tendon sheath may be involved, the knee, hip, and ankle are the preferred targets [3–5].

Tenosynovial D-GCT has been reported to recur in 10% to 45% of cases [6]. Metastases to the lymph nodes or lung may develop [7–9]. We report the case of a patient with lung metastases from a D-GCT of the fibular tendon sheath at the ankle. This case highlights the importance of close monitoring of these tumours reported to be only locally aggressive and the usefulness of positron-emission tomography (PET) monitoring in patients with recurrent D-GCT.

2. Case report

A 56-year-old male farmer had surgery for a GCT of the fibular tendon sheath at the right ankle. The resection margin was R1. He was referred to us 2 years later, in June 2008, for a recurrence. Magnetic resonance imaging (MRI) visualised a soft-tissue growth behind and about the right lateral malleolus, at the level of the tibio-tarsal joint. Post-gadolinium enhancement was heterogeneous. The joint was not involved. No metastases were seen by PET. Six Tru-cut biopsies (14 G) confirmed the diagnosis of tenosynovial D-GCT by showing soft-tissue infiltration by mononuclear cells, giant multinucleated cells, and hemosiderin deposits (Fig. 1). By immunohistochemistry, both the mononuclear cells and the giant multinucleated cells were positive for the enzyme phospho-gluco-mutase 1 (PGM1). Surgical excision was selected as the best treatment option during a regional multidisciplinary meeting. Resection margins were R1. The histological diagnosis was recurrent tenosynovial D-GCT with no sarcomatous changes. Clinical follow-up was provided and MRI scheduled 6 months after surgery.

In March 2009, clinical abnormalities consistent with a second recurrence were found. MRI was performed and ultrasound-guided biopsies discussed during a regional multidisciplinary meeting. A diagnosis of recurrent D-GCT was given. PET detected four new foci...
at the ankle and right leg, as well as six lung metastases with a maximum standardised uptake value (SUV) of 39 (Figs. 2 and 3). A lung biopsy (Fig. 4) confirmed the diagnosis of metastatic lung disease from the D-GCT (Fig. 5). The treatment strategy selected during a supra-regional multidisciplinary meeting was targeted therapy with imatinib, radical treatment of the primary and leg lesions by amputation, surgical excision of the right lung tumours, and radiofrequency ablation of the left lung tumours. Fatal pulmonary embolism occurred within a few days after the thoracic surgical procedure. The pathological study of the amputation specimen found a 9-cm by 16-cm primary tumour and four other intra-muscular tumours measuring 1.4 cm to 2.5 cm along their greatest diameter with tumour emboli. The pathological findings confirmed the diagnosis of lung metastases from the known D-GCT, without evidence of sarcomatous changes.

3. Discussion

Tenosynovial GCT presents clinically as a slow-growing painless nodule of fibro-elastic consistency that adheres to the underlying tissues. The surrounding tissue is extensively destroyed and a radiographic erosion indicating extension to neighbouring bone is visible in 10% to 20% of cases [2,6,10]. MRI is the investigation of choice for guiding the diagnosis [11]. The tumour generates low signal on T1-weighted images and high signal on T2-weighted images. Nevertheless, the hemosiderin deposits and abundant collagen matrix cause a typical decrease in signal intensity, which is most noticeable on the T2-weighted sequence [11]. These features distinguish tenosynovial GCT from other solid tumours of the soft-tissues [12]. The definitive diagnosis requires a histological examination. The cell population, of variable density, combines mononuclear cells, giant multinucleated cells, and xanthomatous cells. The tumour also contains a variable number of hemosiderin deposits [3].

Local GCT recurrences (10%–45% of cases) are often due to incomplete resection, with R1 or R2 margins [8]. The treatment of choice for GCT is therefore complete resection (R0 margins) [9]. Adjunctive radiotherapy has been suggested in patients at high-risk for recurrence but is very rarely used given the associated risk of secondary malignant transformation [13]. Imatinib therapy deserves consideration in patients with uncontrollable or inoperable D-GCT. The rationale for imatinib therapy lies in the overexpression by GCTs of colony-stimulating factor 1, which is a chemoattractant for the inflammatory cells.
that constitute most of the tumour population. Imatinib is a

tyrosine kinase inhibitor that targets colony-stimulating factor 1

[14,15].

Metastases can develop from tenosynovial GCTs, including

those with unremarkable histological features. Dissemination

may occur when tumour emboli are mobilised during surgical

manipulation of the tumour [9]. Three cases of lung or lymph

node metastases from tenosynovial GCTs have been reported

[15,16].

4. Conclusion

Tenosynovial GCTs should be excised as extensively as possible,

ideally with R0 margins. Regular follow-up evaluations are

mandatory and MRI is advisable 6 months after surgery. The rou-
tine evaluation of local recurrences should include MRI and a biopsy

examined at a referral centre to look for sarcomatous transforma-
tion. In patients with diffuse and/or recurrent GCTs, PET is useful
to look for node or lung metastases.
Disclosure of interest

The authors declare that they have no competing interest.

References