Rare disease

Rare sarcoma presented as sinusitis

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Summary

Myxofibrosarcoma (MFS) also known as myxoid variant of malignant fibrous histiocytoma is one of the most common soft tissue sarcomas of the extremities in adult and elderly patients with rare occurrences in head and neck region. Low-grade MFS is unusual among low-grade sarcomas because it often recurs relentlessly and multiplies despite wide local resection with gross negative margins. We report a case of extreme rarity and a tumour of aggressive nature in the maxillary sinus, which presented with non-specific sinonasal symptoms and we present a review of the radiological and histopathological characteristics of this rare tumour and recent evidence of management.

BACKGROUND

Low-grade myxofibrosarcoma (LGMFS) is unusual among low-grade sarcomas because it often recurs relentlessly and multiplies after surgical resection, despite gross negative margins and wide surgical resection.

We reported a case of extreme rarity, a tumour of aggressive nature in the maxillary sinus, which presented with non-specific sinonasal symptoms. Awareness and high index of suspicion among clinicians is vital. Early detection of these fatal tumours can change the morbidity of the surgery and attain the patient remission and excellent prognosis. The diagnosis of sinusitis is mainly clinical and straightforward (nasal congestion/blockage, rhinorrhoea/postnasal drip, smell disorder and facial pain/discomfort) and any unusual presentation or atypical symptoms such as severe or prolonged facial pain, numbness or hyperesthesia, loose denture, double vision, proptosis and epistaxis must draw the attention of a clinician to more sinister possibility.

CASE PRESENTATION

A 50-year-old lady of middle-eastern origin presented to our department with a 2-month history of left chronic sinus pain associated with left-sided toothache. Initial assessment showed a minimal symptomatic relief despite the use of repeated courses of antibiotic, analgesia and topical nasal steroids prescribed earlier by her doctor and local dentist. On examination, a mild proptosis in the left eye was noticed associated with a 3 mm inferior scleral show in upper gaze with diplopia. Endoscopic nasal examination was unremarkable. A full otolaryngological examination was normal.

INVESTIGATIONS

An urgent CT scan of the paranasal sinuses illustrated a large mass of mixed attenuation and enhancement filling the left maxillary sinus with expansion into left inferior orbital wall. The mass had eroded superiorly into the lamina papyracea and the posterolateral wall of the sinus. There was also erosion of the left maxillary alveolus, which could explain for her dental complaint (figure 1).

An MRI scan confirmed the findings of the CT scan and excluded both pterygopalatine soft tissue extension and orbital periosteum involvement (figure 2). CT imaging of the neck and chest were both clear. Haematology and chemistry studies were all normal. Endoscopic examination under anaesthesia later on showed bowing of the lateral nasal wall in the inferior meatus with a normal looking middle meatus (figures 3 and 4); hence, several biopsies taken from inferior turbinate, meatus and maxillary sinus through the inferior antrostomy.

Diagnostic histology featured spindle-shaped and stellate cells with poorly defined cytoplasm and pleomorphic nuclei, in a myxoid stroma. There were thin-walled curvilinear vessels within the tumour. The mitotic index is 1–2/10 high-power field. Necrosis was not identified. Scattered lymphocytes, plasma cells and mast cells were noted. Immunostaining showed that the spindle cells did

Figure 1 Coronal CT section.

Figure 2 MRI scan T1 axial section.

not express DESMIN, SMA, EMA, CD68 or S100. Scattered CD68 positive monocytes/macrophages cells were present (figures 5 and 6).

DIFFERENTIAL DIAGNOSIS

- ▶ Sinonasal tumour
- ▶ Rhinosinusitis
- ▶ Nasal polyps
- ▶ Teeth root retention
- ▶ Dental infection
- ▶ Silent sinus syndrome

TREATMENT

The lesion was excised en-bloc (maxillectomy with orbital preservation) via midfacial degloving approach and all margins were clear of the disease. Patient was referred afterwards for adjuvant radiotherapy. However, in last December, the patient had another craniofacial procedure to extirpate recurrent disease.

Figure 3 Endoscopic view of middle meatus.

Figure 4 Endoscopic view of the left inferior meatus.

OUTCOME AND FOLLOW-UP

The recovery with each procedure was uneventful and she currently remains on regular review under a multidisciplinary team as part of head and neck cancer surveillance monitoring.

DISCUSSION

LGMFS is slow growing and rarely leads to distant metastases. But the local recurrence rate of the low-grade type is as high (50–60%) as that of the high-grade type. The middle-aged and elderly are most commonly affected (1)¹ with an approximately equal incidence in men and women.² Regions affected by the tumour as published in various series showed that the most common site was the lower limb (57%), followed by the upper limb (31%), trunk (8%) and lastly head and neck (4%).³

LGMFS in the sinonasal tract usually exhibits features similar to other soft tissue neoplasms, namely epistaxis, rhinorrhea, nasal obstruction and non-specific nasal discomfort. However, there are no distinguishing physical signs to LGMFS.⁴

Figure 5 Macroscopic view.

Figure 6 Microscopic view.

Findings pertain to the relatively smooth surface of the tumour and its uniform density or no necrotic area; the clear separation of the tumour from the surrounding soft tissues on the CT images.⁵ A poor periosteal reaction to the tumour invasion; that is an osteolytic bone change was also noticed in some cases.

LGMFS showed a myxoid component, which is defined on MRI as a lesion showing hypointense T1-weighted and extrahyperintense T2-weighted signals. The presence of an increased perifacial T2 signal is characteristic in myxofibrosarcoma. Fluid level might be present in some cases.⁶ The infiltrative growth pattern may be obvious in gadolinium-enhanced fat-suppressed T1-weighted images even in tumour with focal growth pattern. In addition, diffusion-weighted imaging can improve assessment of infiltration with myxofibrosarcoma.⁶

Tumour cells of myxofibrosarcoma are likely to be derived from fibroblastic and primitive mesenchymal cells rather than from a histiocytic lineage.⁷

Myxofibrosarcoma were graded according to the system proposed by Mentzel *et al.*⁷ All tumours showed a broad spectrum of histological grade characterised as low, intermediate and high. The principle feature shared by all tumours of any grade was a varying proportion of hypocellular myxoid and cellular areas, the myxoid areas being more prominent in lower-grade lesions.

Macroscopically (figure 5) the resected tumour is very soft and well circumscribed. Its cut surface is uniformly gelatinous and yellow-grey in colour.⁸ Microscopically (figure 6), myxofibrosarcoma comprises a continuous range from hypocellular tumours, consisting of small spindle cells widely spaced among the myxoid matrix, to more cellular variants with pleomorphic nuclei and pseudolipoblasts.

LGMFS must be distinguished from other benign and malignant mesenchymal lesions with a prominent myxomatous matrix. It is also prone to misinterpretation as other benign or malignant myxoid mimickers such as myxoid liposarcomas, Low-grade fibromyxoid sarcomas (FMSs), cellular myxomas, nodular fasciitis, myxoid neurofibroma, myxoma and superficial angiomyxoma.^{4–8} LGMFS and low-grade FMS are two distinct entities in the spectrum of myxoid mesenchymal sarcomas with fibroblastic differentiation. Low-grade FMS is characterised by an indolent but ultimately malignant clinical course with metastases in

more than half of the cases, which underlines the importance of distinguishing between the two entities.

The capacity of lower-grade lesions to advance in histological grade underpins the necessity for accurate diagnosis and adequate excision of these lesions, especially because tumours that are purely low grade seem to have little or no potential for metastasis and are therefore potentially curable. LGMFS demonstrates a high rate of local recurrence if treated inappropriately. Low-grade lesions often become higher grade in subsequent recurrence and acquire metastatic potential. Therefore, tumour resection with wide surgical margin is critical for disease cure.⁶ However, achieving a complete resection with wide margins is generally not possible in the head and neck region.

Adjuvant radiotherapy is usually reserved for recurrent, unresectable or large lesion with high chance of microscopic tumour involvement of the margin, its efficacy is controversial.⁴ Fayda *et al.*⁹ in their study showed that there is no difference in local control between patients with negative or microscopically positive disease if they receive postoperative radiotherapy. Regarding overall survival, median survivals of patients receiving both surgery and radiotherapy with negative and microscopically positive margins were significantly better than patients who were not treated with surgery.⁹ Therefore, the treatment strategy is a combination of surgical excision and radiotherapy. The goal of surgery is to obtain tumour-free margins of 1–3 cm.^{10–11}

Chemotherapy has been used for the treatment of myxofibrosarcoma, but the results are poor. Likewise, the use of adjuvant chemotherapy does not significantly affect local control. Zagars *et al.*¹⁰ found no evidence that adjuvant chemotherapy decreased the metastatic or local recurrence rates.

The overall 5-year and 10-year survival rates for LGMFS are 65% and 52%, respectively.¹² Patient age, tumour size (>5 cm), histological grade, positive section margin and deep-seated tumours are all poor prognostic factors for various soft tissue sarcomas of the head and neck and significantly associated with poor overall survival.^{13–14}

Learning points

- ▶ Rare and potentially fatal tumours can give few or no symptoms in clinical context; hence, high index of suspicion is very important in all cases of unusual presentation or failure of responding to empiric treatment.
- ▶ Sinonasal pain is unusual in cases of chronic sinusitis. Patients with assumed sinus infection (nasal congestion/blockage, rhinorrhoea/postnasal drip, smell disorder and facial pain) should be diagnosed and treated according to the sinusitis protocol. Any unresponsiveness or failure should be dealt with by referring the patient to the specialist.
- ▶ Rare sarcoma proved to be difficult to diagnosis clinically, radiologically and histologically.
- ▶ Low-grade myxofibrosarcoma is rare and very aggressive tumour, hence early intervention is important.
- ▶ Early referral to the specialist is necessary, if the clinical presentation is unusual.

Competing interests None.

Patient consent Obtained.

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