CASE REPORT

Malignant fibrous histiocytoma

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SUMMARY

Malignant fibrous histiocytoma (MFH) is a type of histiocytoma and is the most common soft tissue sarcoma of late adult life. However, it is relatively uncommon in the head and neck area. It usually occurs/develops in the lower extremities and in the retroperitoneum. This tumour is difficult to distinguish histologically from other sarcomas and carcinomas. Surgery is the only treatment option. Prognosis is fairly poor; recurrence and local metastasis are common. In comparison with MFH of the extremities and the trunk, the 5-year survival rate for cases of this tumour in the head and neck is low. It is important to consider MFH in differential diagnosis of head and neck tumours because of its poor prognosis. We report a rare case of MFH in an 11-year-old girl.

BACKGROUND

Malignant fibrous histiocytoma (MFH), the most frequent soft tissue sarcoma of adulthood, was first described as a new malignant tumour by O'Brian and Stout in the 1960s¹ and the details of the histopathological features of MFH were first described by Kempson and Kyriakos.² MFH of bone was first described by Feldman and Norman in 1972.³ MFH arises predominantly in soft tissue and is one of the most common malignant tumours

among elderly. MFH occurs most commonly in the extremities (70–75%) with lower extremities accounting for 59% of cases, followed by the retroperitoneum. Tumours typically arise in deep fascia or skeletal muscle. MFH has been reported to occur in the lung, kidney, bladder, scrotum, vas deferens, heart, aorta, stomach, small intestine, orbit, central nervous system, paraspinal area, duramater, facial sinuses, nasal cavity and oral cavity.⁴

In the head and neck, the nasal cavity and the paranasal sinuses are the most commonly affected sites (54.3%), and can lead to subsequent involvement of the maxillary alveolar bone. Larynx, maxillary sinus and mandible have the worst prognosis in the head and neck lesion.⁵

CASE PRESENTATION

A 11-year-old girl presented with a 2-month history of rapidly increasing swelling over right lower back gingival region. On examination, the patient was a healthy individual with normal vital signs (figure 1). The patient's medical history was unremarkable. On local examination, a localised reddish bilobulated swelling was present on the right lower gingival molar region measuring approximately 2×2 cm in diameter with smooth overlying surface (figure 2).

The mass had implicated the alveolar ridge buccolingually and extended to the adjacent mandibular vestibule. Right deciduous first and second mandibular teeth were mobile. The mass had a soft to firm consistency in some areas. It was mostly of firm consistency, without any reported tenderness or paraesthesia.

Figure 1 Profile photograph of the patient.

Figure 2 Intraoral examination revealed a localised reddish bilobulated swelling was present on the right lower gingival in molar region measuring approximately 2×2 cm in diameter with smooth overlying surface.

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Figure 3 Panoramic radiograph showed marked irregular bony erosion in the involved area with no displacement and resorption of teeth.

INVESTIGATIONS

Panoramic radiograph showed marked irregular bony erosion in the involved area with no displacement and resorption of teeth (figure 3).

Mandibular cross-sectional occlusal radiograph did not show any bony expansion (figure 4).

For confirmation an incisional biopsy was performed under local anaesthesia and the obtained tissue was sent for routine histopathological examination (figure 5).

Histopathology showed spindle-shaped fibroblast arranged in storiform pattern around the slit-like vessels along with the presence of the histiocytes cells. Malignant cells are round to ovoid with nuclei pleomorphism and atypical mitotic figures (figure 6).

DIFFERENTIAL DIAGNOSIS

- ▶ Giant cell tumour of the mandible
- ▶ Peripheral giant cell granuloma
- ▶ Giant cell tumour secondary to hyperparathyroidism
- ▶ Fibrosarcoma of the mandible
- ▶ Other sarcomas and soft tissue tumours
 - Synovial sarcoma
 - Aggressive fibromatosis
 - Benign fibrous tumours
- ▶ Soft tissue metastases
- ▶ Myositis ossificans

TREATMENT

The patient was referred to the department of oral and maxillo-facial surgery for the treatment of growing mass.

Figure 4 Mandibular cross-sectional occlusal radiograph did not showed any bony expansion.

Figure 5 Incised tumour specimen.

Surgery was performed because of anatomical limitation and as the patient was young.

OUTCOME AND FOLLOW-UP

The prognosis for the current case was poor.

DISCUSSION

The term 'MFH' or 'pleomorphic sarcoma' was first introduced in the early 1960s to refer to a group of soft-tissue tumours, characterised by a storiform or cartwheel-like growth pattern.⁶ MFH is classified into primary and secondary types. Seventy per cent of MFH are primary tumours that involve younger patients while the secondary tumours are seen in sixth and seventh decades of life. The secondary tumours are more aggressive than the primary ones. They are associated with an underlying condition such as Paget's disease, fibrous dysplasia or prior radiotherapy.⁷

The biological behaviour of MFH shows a high incidence of local recurrence and metastasis and carries a poor prognosis.

Figure 6 Histopathologically, examination with H&E stain showed spindle-shaped fibroblast arranged in storiform pattern around the slit-like vessels along with presence of the histiocytes cells. Malignant cells are round to ovoid with nuclei pleomorphism and atypical mitotic figures.

The lungs are the most common site of metastases from MFHs arising in the head and neck.⁸

According to Huvos, metastatic spread in patients with MFH primarily in bone occurs via haematogenous dissemination, predominantly in the lungs, rather than in regional lymph nodes.

Based on cellularity, pleomorphism, necrosis and mitotic activity, MFH is classified as low grade for well differentiated, intermediate grade for moderate differentiated and high grade for poorly differentiated and undifferentiated. Based on histology MFH is classified into five types—storiform/pleomorphic, giant cell, inflammatory, myxoid (myxofibrosarcoma) and angiomatoid.⁹ Pleomorphic is the most commonly seen histological variant of tumour and our case also belonged to this subtype.

According to review carried out by Kanazawa, among the MFH cases, 65% men and 35% women showed a distinct gender predilection. The age distribution at diagnosis varied from 1.5 to 69 years, with a mean age of 41. The most common presenting symptom of MFH was a painless, gradually progressing mass without any mucosal ulceration, mandibular lesions were usually first noticed by swelling, pain, paraesthesia and loosening of teeth.¹⁰ In the case of our patient painless progressing mass there was no pain and paraesthesia but loosening of tooth was present. Kanazawa *et al* reported that lesions of the mandible presented as an extensive, ill-defined osteolytic lesion radiographically and neither marginal sclerosis nor periosteal reaction was observed; similar features were evident in our patient also. Xuexi *et al* reported that the extent of surgery may be defined as radical, wide or local resection. In the treatment protocol, radical surgery with a minimum margin of 3 cm is the choice of treatment followed by the combination of radiotherapy and/or chemotherapy as advocated by most of the authors.¹¹ MFH is generally treated the same way as osteosarcoma and appears to have a similar response to treatment. MFH treatment options vary based on factors such as: (1) whether the MFH is localised or metastatic, (2) the location of the tumour and (3) the patient's age and general health. In general, treatment options for MFH may involve surgery and chemotherapy. Patients should coordinate with their doctors to develop a treatment plan that meets their medical needs and personal values.^{5 7 8} Choosing the most appropriate MFH treatments is a decision that ideally involves the patient, the family and the healthcare team. Treatment of localised MFH may include chemotherapy followed by surgery to remove the tumour and surrounding tissue.^{7 9 11} There is no standard treatment for cancer that cannot be removed with surgery or cancer cells that

have spread (metastasised). In these situations, the MFH prognosis is poor.^{10 11}

Learning points

- ▶ Head neck malignant fibrous histiocytoma is rare but aggressive with poor prognosis. Radical resection with adjuvant chemotherapy is the treatment of choice.
- ▶ Local radical resection was performed along with primary reconstruction by temporalis muscle and local buccal fat, followed by adjuvant chemotherapy.
- ▶ Prognosis of malignant fibrous histiocytoma lesions is influenced by the depth of tumour infiltration into the surrounding tissue, the mass of the tumour and anatomic location of the tumours.

Competing interests None.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

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