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Clinical Record

Myxofibrosarcoma of the neck

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Abstract

We report a rare case of myxofibrosarcoma arising in the neck. A 55-year-old man presented with a two-year history of left-sided, painless, submandibular swelling. Computed tomography and magnetic resonance imaging (MRI) revealed an 80 × 35 mm, well defined, lobulated, submandibular tumour extending to the parapharyngeal space. The tumour showed uniformly low intensity and marked hyperintensity in T1- and T2-weighted MRI scans, respectively, and was scarcely enhanced by gadolinium. A tentative diagnosis of lymphangioma or plunging ranula was made, and the patient underwent local injection of OK-432, which proved to be ineffective. Resection of the tumour was then performed via a transcervical approach. The tumour was histopathologically and immunohistochemically diagnosed as a low-grade myxofibrosarcoma. The patient's post-operative clinical course was uneventful, and the patient was free of disease 27 months after surgery. The pathology, clinical characteristics and treatment of myxofibrosarcoma are bibliographically reviewed.

Key words: Myxofibrosarcoma; Malignant Fibrous Histiocytoma; Neck

Introduction

Myxofibrosarcoma, also known as a myxoid variant of malignant fibrous histiocytoma (MFH), is a fibroblast-derived soft tissue neoplasm usually arising in late adulthood. The most common locations are the lower and upper limbs, with rare occurrences in the head and neck.[1] To our knowledge, only two cases in the neck have so far been documented.[2,3]

We report an additional case of myxofibrosarcoma, arising in the submandibular region, and review the clinical, radiological and histopathological characteristics of this tumour.

Case report

A 55-year-old man had noticed a painless, left-sided, submandibular swelling which had gradually increased in size over the past two years. When he was first seen by us in July 2003, a fist-sized, very soft and non-tender mass was present in his left submandibular region. Otorhinolaryngological findings were otherwise normal, and no cervical lymph nodes were palpable. Chest X-ray and laboratory data were unremarkable. The patient had no particular past or family history of illness.

Computed tomography and magnetic resonance imaging (MRI) revealed an 80 × 35 mm, well defined, lobulated, submandibular tumour extending to the parapharyngeal space (Figure 1). The tumour showed uniformly low intensity and marked hyperintensity in T1- and T2-weighted MRI scans, respectively, and was scarcely enhanced by gadolinium.

Under a tentative diagnosis of lymphangioma or plunging ranula, the patient underwent local injection with OK-432 (5KE), which proved to be ineffective. Resection of the tumour via a transcervical approach was then performed in December 2003. Adjoining tissues, such as the submandibular gland, marginal mandibular branch of the facial nerve, and digastric and mylohyoid muscles, were uninvolved and were easily separated from the tumour. No connection to the submandibular or sublingual glands was seen. The deep end of the parapharyngeal portion of the tumour was carefully separated from surrounding tissue by blunt digital dissection.

Macroscopically, the resected tumour was very soft and was circumscribed. Its cut surface was uniformly gelatinous and yellow-grey in colour. Microscopically, the tumour consisted of hypocellular to moderately cellular myxoid areas, with tumour cells varying from small and bland to enlarged, bizarre, pleomorphic and multinucleated. The cytoplasm of the cells was scant and eosinophilic, and mitoses were infrequent. Immunohistochemically, the tumour cells were positive for vimentin and CD34, while stains for S-100 protein, cytokeratin, desmin and α-smooth muscle actin were negative.

On the basis of these histopathological and immunohistochemical findings, we diagnosed the tumour as a low-grade myxofibrosarcoma. Because the surgical margin was microscopically free of tumour, no additional treatment was given to this patient.

The patient's post-operative clinical course was uneventful. He was discharged on the eighth post-operative day and was free from disease 27 months after surgery.

Discussion

In 1977, Weiss and Enzinger[4] reported a variant of MFH that exhibited a highly myxoid, hypocellular appearance and had a better prognosis than usual, nonmyxoid forms

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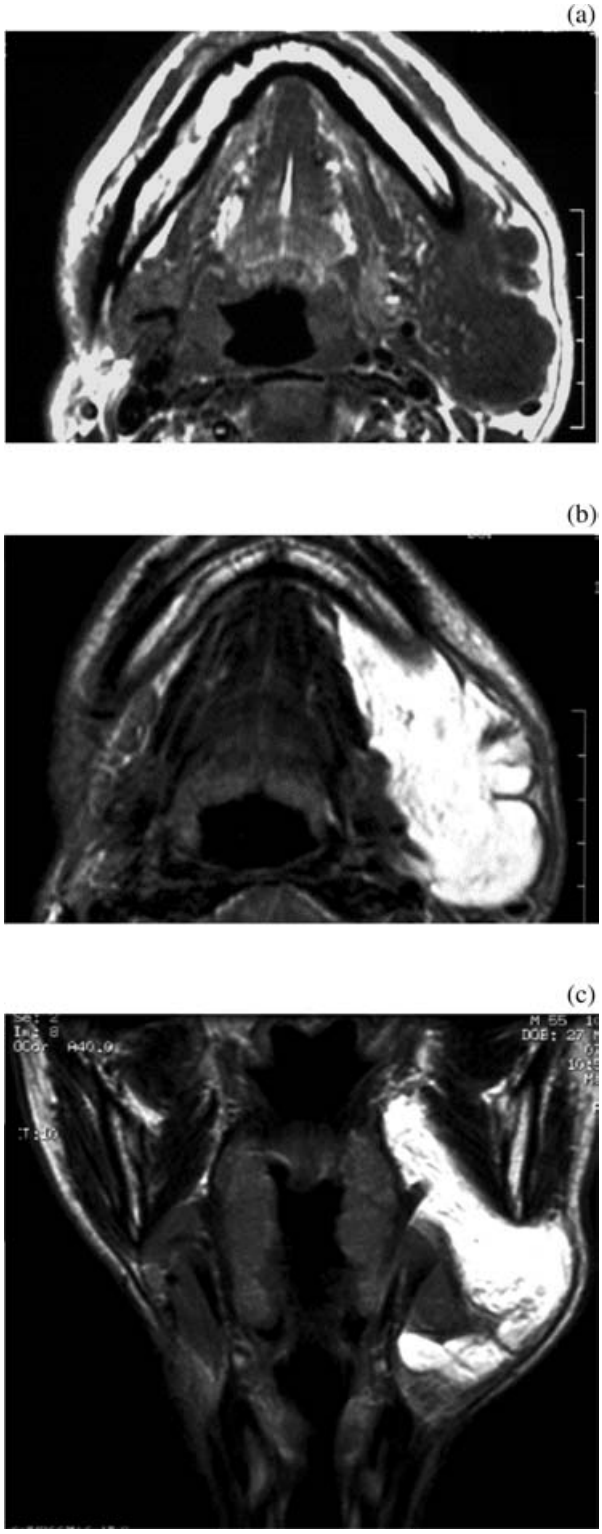

FIG. 1

Magnetic resonance imaging (MRI) scans showing an 80 × 35 mm, well defined, lobulated, submandibular tumour extending to the parapharyngeal space. (a) Axial T1-weighted MRI; (b) axial T2-weighted MRI; (c) coronal T2-weighted MRI.

of the tumour. This myxoid variant of MFH was comprehensively studied in the light of modern histopathology by Merck *et al.*,[1] who referred to it as myxofibrosarcoma. Later immunohistochemical studies have shown that the

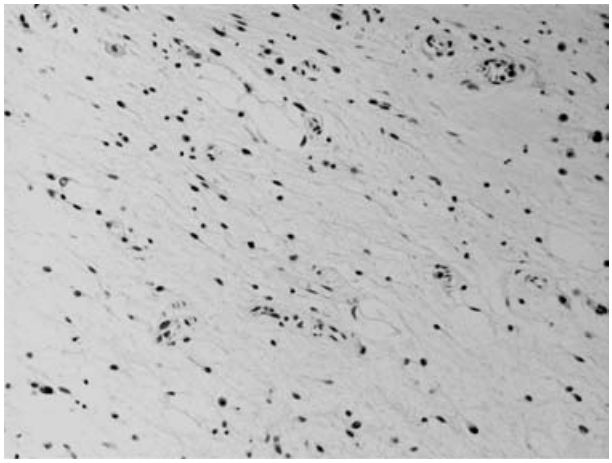

FIG. 2

Photomicrograph of the tumour. The tumour comprises a hypocellular to moderately cellular myxoid lesion with tumour cells varying from small and bland to enlarged, bizarre, pleomorphic and multinucleated. The cytoplasm of the cells is scant and eosinophilic, and mitoses are infrequent (H&E; ×200).

tumour cells of myxofibrosarcoma are likely to be derived from fibroblastic and primitive mesenchymal cells rather than from a histiocytic lineage.[5] This tumour predominantly occurs in the lower and upper limbs, most often in the thigh, of elderly people. A previous study documented a slight male predominance of myxofibrosarcoma,[1] whereas another author reported that this tumour equally affects both sexes.[5]

Histologically, myxofibrosarcoma may exhibit various proportions of myxoid matrix with varied cellularity. Merck *et al.*[1] categorized myxofibrosarcomas into grades I to IV according to the degree of tumour cellularity, cellular atypia and prevalence of mitotic figures. More recently, Mentzel *et al.*[5] divided the lesions into low, intermediate and high grades to simplify the Merck classification system.

Low-grade lesions show hypocellular to moderately cellular architecture with a prominent myxoid matrix. Tumour cells are fusiform, round or stellate, with hyperchromatic and irregularly shaped nuclei, and they assume mild pleomorphism. Mitoses are seen only occasionally.

In contrast, high-grade lesions show more solid and hypercellular architecture with small myxoid areas. Cellular pleomorphism and mitotic figures are conspicuous. Areas of haemorrhage and necrosis are also observed.

Intermediate-grade lesions lack areas of necrosis and pronounced cellular pleomorphism, but are more hypercellular and manifest a higher mitotic count than do low-grade lesions.

- **Myxofibrosarcoma is a variant of malignant fibrous histiocytoma**
- **A rare case of this tumour arising in the neck is reported**
- **The pathology, clinical characteristics and treatment are reviewed**

Immunohistochemically, the tumour cells of myxofibrosarcoma are generally positive for vimentin and negative for S-100 protein, CD68 and desmin.[5] Some high-grade lesions may express muscle-specific actin and smooth

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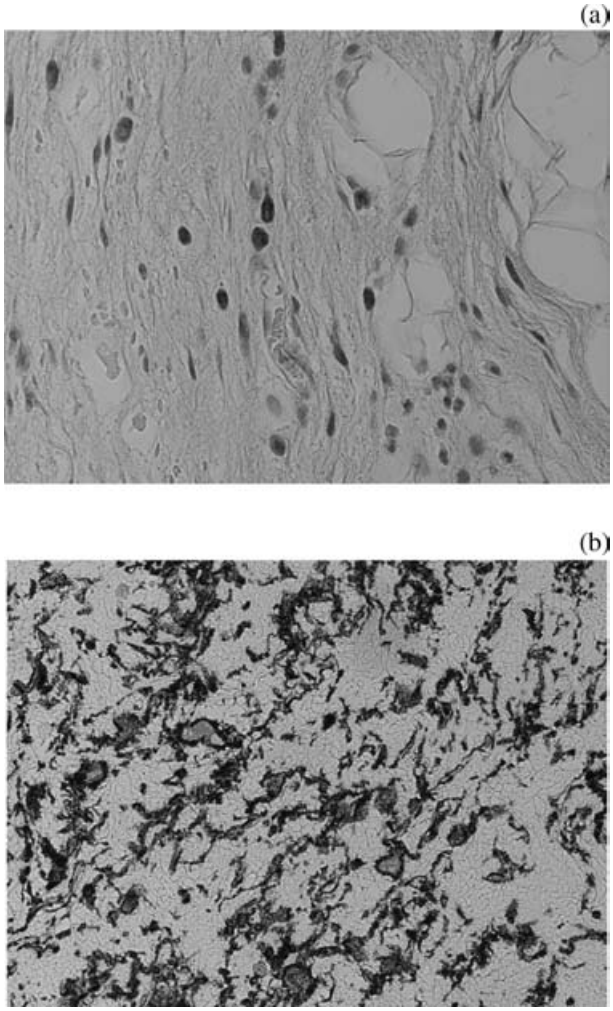

FIG. 3

Immunohistochemical findings. The tumour cells are positive for (a) vimentin and (b) CD34. Note intense and membranous immunoreactivity for CD34 in many spindle cells (immunoperoxidase technique; ×400).

muscle actin, suggesting myofibroblastic differentiation.[5] The histopathological findings of the present case were compatible with a diagnosis of low-grade myxofibrosarcoma, and the immunoreactivity to vimentin and CD34 suggested the primitive fibroblastic nature of the tumour cells.[6]

Low-grade myxofibrosarcoma may sometimes be confused with benign myxomatous tumours such as myxoid neurofibroma, myxoma and superficial angiomyxoma. Myxoid liposarcoma is a common low-grade malignant sarcoma and is difficult to distinguish from myxofibrosarcoma. Atypical lipoblasts and a thin-walled capillary network organized in a plexiform pattern are characteristic of myxoid liposarcoma.[1,7] The differential diagnosis also includes a more aggressive malignancy, fibromyxoid sarcoma, which occurs in younger patients and metastasizes more frequently than does myxofibrosarcoma.[5] Such tumours can be distinguished from myxofibrosarcoma by a careful histopathological examination.

Complete excision with a tumour-free surgical margin is the only curative treatment for myxofibrosarcoma.[1,4,5] There are a limited number of reports on radiotherapy and chemotherapy for this disease; neither therapy is effective.[1] The local recurrence rate is reported to be 50 to 61 per cent[1,4,5] and is independent of the grade of malignancy.[5] The overall five- and 10-year survival rates are 65 and 52 per cent, respectively.[1] According to Merck *et al.*, the five-year survival rate for patients with grade I or II lesions is more than 80 per cent, whereas that for patients with grade IV lesions is less than 50 per cent.[1] The incidence of distant metastasis is 20 to 24 per cent.[1,4,5] Mentzel *et al.* documented that none of the low-grade myxofibrosarcomas in their study metastasized; most of the metastasizing lesions were high-grade in their study.[5] These lines of evidence suggest that low-grade myxofibrosarcomas have negligible metastatic potential, implying a good short-term prognosis. However, we must be aware that this tumour frequently recurs and may become progressively more malignant as it does so.[5] Therefore, these patients should be placed under careful and long-term follow up.

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