Malignant Fibrous Histiocytoma: An Uncommon Sarcoma with Pathological Fracture of Mandible

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Received: 22 October 2012 / Accepted: 19 February 2013 / Published online: 27 March 2013
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Abstract

Background Tumors composed of cells differentiating as both fibroblasts and histiocytes have been designated fibrous histiocytomas. Only a small percentage of these lesions behave in a malignant fashion, they are called malignant fibrous histiocytoma (MFH). The occurrence of MFH in membranous bones including the mandible is quite unusual. Involvement of the mandible accounts for only 3 % of all MFH bone lesions. Recent literature revealed only a few (30) cases of MFH involving the mandible but not a single case of MFH associated with pathological fracture of the mandible, probably the first such kind of case to support antecedent trauma as an initial proliferative response for its occurrence.

Case report A rare case of MFH involving the mandible and submandibular glands with pathological fracture in a 14-year-old boy is presented with special emphasis on the poor prognosis even after prompt therapy, its controversial histogenesis, high malignant potential, high recurrence rate and tendency to metastasise. The patient was regularly followed up for 11 months post-operatively, during which patient complained of pain in the operated region after 8 to 9 months. Though there was no clinically obvious abnormality seen, the young boy died after 11 months. The clinical, surgical, radiographic and pathological features of this lesion are discussed.

Discussion Malignant fibrous histiocytoma, the most frequent soft tissue sarcoma of adulthood, was first described as a new malignant tumour by O'Brian and Stout in the 1960s and the details of the histopathological features of MFH were first described by Kempson and Kyriakos. Despite the frequency of diagnosis, MFH has remained an enigma as no true cell of origin has ever been identified. Treatment consists of surgical excision and in some cases chemotherapy and radiation. Early and complete surgical removal using wide or radical resection is indicated because of the aggressive nature of the tumor. The combination of infrequent occurrence, varied pathologic features, uncertain histogenesis, numerous subtypes and the many potential sites of presentation makes these tumors a challenge for the diagnostician, surgeon and oncologist. Close follow-up after treatment is important, as local recurrence is common and early metastasis to the lungs is also frequent, which are the reasons for high mortality rate in MFH.

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Keywords Malignant fibrous histiocytoma · Jaw neoplasms · Uncommon sarcoma · Pathological fracture · Schwannoma of maxilla · Plexiform schwannoma · Intraosseous schwannoma · Neurilemmoma

Background

Tumors composed of cells differentiating as both fibroblasts and histiocytes have been designated fibrous histiocytomas. Only a small percentage of these lesions behave in a malignant fashion, they are called malignant fibrous histiocytoma (MFH) [1]. Head and neck MFH have been reported to account for 3–10 % of MFH formed in various parts of the body. The occurrence of MFH in membranous bones including the mandible is quite unusual. Involvement of the mandible accounts only for 3 % of all MFH bone lesions [2]. Malignant fibrous histiocytomas are most commonly found in the extremities. The commonest site of MFH are the soft tissues of the limbs and retroperitoneum, and only limited numbers have been documented in the jaws and oral cavity [3]. Approximately 70 % of MFHs arise as primary tumors while the other 30 % are associated with pre-existing conditions such as prior radiation therapy to the region [2].

MFH arises predominantly in soft tissues, and is one of the most common malignant tumours among the elderly, with a peak incidence in the fifth to the seventh decades of life. Also, it is the most common malignant soft tissue sarcoma of this age group. In rare cases MFH does occur in children, but it is usually in a less aggressive form [4]. In the 1970s, Feldman and Norman described the first primary tumor of bone that satisfied the histological criteria of MFH. 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].

We present a rare case of MFH involving the mandible in a 14-year-old boy. This case presented the opportunity to analyze the clinical aspects of previously reported mandibular intraosseous MFH.

Case Report

A 14 year old boy presented with facial asymmetry, diffuse extra oral swelling over the right side of mandible since 1 year. History of trauma because of a fall from the bicycle 2 years back and repeated trauma to the jaw a year back was elicited. Intraorally the swelling was over the mucovestibule, floor of the mouth in premolar-molar region on the right side of the mandible which was nonmovable, hard, painless and was gradually increasing in size. There was crepitus in left parasymphysis region. Regional lymph nodes were palpable, enlarged, movable and tender.

Radiological examination revealed a radiolucent lesion with ill defined border, scattered radiolucency with mixed flakes of radiopacity extending from left canine-premolar region to the right angle of the mandible. Root resorption

of multiple teeth with hanging teeth appearance and pathologic fracture of the mandible in left parasymphysis region was evident. Chest X-ray did not reveal any metastatic deposits. Conclusive diagnosis could not be achieved on incisional biopsy, multiple possibilities like osteosarcoma, osteomyelitis, metastatic carcinoma were suggested.

Resection of mandible with radical neck dissection of was planned. The mandible was exposed from right angle region to left 1st molar region. Surprisingly, the affected mandibular bone had a soft rubber like consistency and could be easily cut with surgical blade without any hard mandibular bony feeling. Segmental resection of the mandible from right angle to the left premolar along with neck node dissection, and curettage of the lesional tissue was done.

Histologically, the tumor mainly consisted of atypical fibroblast-like spindle cells in a storiform or crisscross pattern. In addition, there were numerous pleomorphic, bizarre cells and mononuclear cells resembling histiocyte. Abnormal mitotic figures, necrosis and extensive cellular atypia were also seen. These findings were suggestive of MFH of storiform-pleomorphic type. The patient was referred to RST cancer hospital, Nagpur, India, for chemotherapy and radiotherapy. The patient was regularly followed-up for 11 months. The patient complained of pain in the operated region after 8–9 months. With local recurrence after 5–6 months patient died with no obvious known reason (Fig. 1).

Discussion

Soft tissue sarcomas of the head and neck are rare mesenchymal malignant neoplasms accounting for less than

Fig. 1 Extra-oral view of the patient illustrating swelling of right side of the face

10 % of all soft tissue sarcomas and approximately 1 % of all head and neck neoplasms. Nevertheless, they represent an important group of tumors and are associated with significant morbidity and mortality [6]. Malignant fibrous histiocytoma, the most frequent soft tissue sarcoma of adulthood, was first described as a new malignant tumour by O'Brian and Stout in the 1960s and the details of the histopathological features of MFH were first described by Kempson and Kyriakos [1]. MFH of bone, first described by Feldman and Norman in 1972, may occur at any age, although most cases occur in patients between the fifth and seventh decades of life. The tumor has a predilection for males [5] (Fig. 2).

Despite the frequency of diagnosis, MFH has remained an enigma as no true cell of origin has ever been identified [5]. In 2002, the World Health Organization declassified MFH as a formal diagnostic entity and renamed it as an undifferentiated pleomorphic sarcoma not otherwise specified. This new terminology has been supported by a compelling body of evidence over the last decade to suggest that MFH represents a final common pathway in tumors that undergo progression towards undifferentiation [6]. Additionally, MFH has been associated with hematopoietic diseases such as non-Hodgkin's lymphoma, Hodgkin's lymphoma, multiple myeloma and malignant histiocytosis [5]. A history of antecedent trauma in about 20 % of the cases suggests that some of these tumors may represent an initial proliferative response to trauma [7] which can be applied to present case also. The proportion of local recurrence rate of MFH after initial local excision ranges between 16 and 52 %, and is related to size, depth of invasion and microscopically positive surgical margins. The lungs are the most common site of metastasis from MFHs arising in the head and neck [1] (Fig. 3).

MFH of the mandible is very rare with only thirty documented cases, showing strong tendency for localization in the

Fig. 2 Panoramic radiograph showing an ill-defined radiolucent lesion extending from right angle to left premolar with pathological fracture of the mandible

Fig. 3 Intra-operative surgical view showing fibrosed, soft mandible

posterior portion of the mandible. Only one case involving anterior portion of the mandible and only four cases of sub-mandibular MFH have been reported [2, 8]. In view of these findings from the literature this particular case becomes distinctly unique as it occurred in the anterior part of the mandible with pathological fracture in left parasymphysis region along with involvement of the submandibular glands (Fig. 4).

Metastatic spread in patients with MFH primary in bone occurs via haematogenous dissemination, predominantly to the lungs, rather than to regional lymph nodes [1]. Kanazawa et al. [9] reported that lesions of the mandible presented as an extensive, ill-defined osteolytic lesion radiographically and neither marginal sclerosis nor periosteal reaction was observed. However, these radiographic findings are not specific to MFH and may be observed in squamous cell carcinoma, fibromyxoma, lymphoma, plasmacytoma and other malignant bone tumors [1] (Fig. 5).

Fig. 4 Histopathological appearance of malignant fibrous histiocytoma ($\times 10$)

Fig. 5 High power view showing histiocyte like cells ($\times 40$)

MFH of the jaws has a distinct clinical behaviour exclusive to this location, being very aggressive with a high rate of local recurrences and frequent metastases [10]. The behaviour of MFH is more aggressive than that of fibrosarcoma and osteosarcoma in this site [8]. The most frequent signs and symptoms at the time of presentation are swelling and pain. Symptoms related to possible involvement of dental roots may also occur such as tooth root resorption [1].

Five histologic subtypes have been described: (1) storiform/pleomorphic, (2) myxoid, (3) giant cell, (4) inflammatory, and (5) angiomatoid [4]. The most common type is storiform-pleomorphic. The most favorable prognosis is associated with the angiomatoid and myxoid types of MFH, whereas a poorer prognosis is reported with the giant cell variant. The giant cell variant represents a highly aggressive lesion in regard to local growth, local recurrence, and distant metastasis. The storiform/pleomorphic and inflammatory MFHs fall between these two extremes [6]. It is common for several morphologic patterns to be observed within one lesion. The explanation of the multiple patterns observed for MFH is related to predominance of cellular differentiation [11].

Treatment consists of surgical excision and in some cases chemotherapy and radiation. Early and complete surgical removal using wide or radical resection is indicated because of the aggressive nature of the tumor. Although local control of the primary tumor is critical to successful treatment of both high- and low-grade lesions, the high rate of distant metastases in high-grade tumors supports the role of combined modality therapy. Since regional lymph nodes are involved in 10–18 % of cases consideration should be given to elective neck dissection for patients with advanced-stage disease or evidence of aggressive histopathological features [6].

Adjuvant radiotherapy is generally recommended for high-grade sarcomas, large tumors, close or positive surgical

margins, and certain histologic variants. Systemic chemotherapy is recommended for those tumors with a significant risk of distant metastasis [4]. Chemotherapy has been used in patients with a predictably high risk of pulmonary metastasis [6]. The administration of anticancer agents such as methotrexate, cisplatin, doxorubicin, cyclophosphamide, actinomycin-D, vincristine, dacarbazine, and ifosfamide has been reported in patients with soft tissue sarcoma. However, tumoricidal effectiveness of each drug is limited to a single administration. It is therefore important to select multiagent combination therapy, considering the interaction of these agents [1].

The efficacy of radiotherapy and chemotherapy treatments for MFH of mandible is not yet well established [9, 10]. The clinical stage of the tumor, which is defined by tumor grade, size, and presence of distant metastases, is the most important prognostic factor [4]. The combination of infrequent occurrence, varied pathologic features, uncertain histogenesis, numerous subtypes and the many potential sites of presentation makes these tumors a challenge for the diagnostician, surgeon and oncologist. Close follow-up after treatment is important, as local recurrence is common and early metastasis to the lungs is also frequent.

Conclusion

MFH, though considered to be the most common sarcoma of soft tissues, can also arise as a primary tumor in bone. We present a rare case of MFH involving mandible in a 14-year-old boy. Recent literature revealed only a few (30) cases of MFH involving the mandible and only four cases having involvement with submandibular glands. Not a single case of MFH associated with pathological fracture of the mandible has been reported in literature, involving submandibular glands also, probably the first such kind of case to support antecedent trauma to be one of the etiological factors, which occurred as an initial proliferative response for Malignant Fibrous Histiocytoma in our case.

Conflict of interest None.

References

1. Senel FC, Bektas D, Caylan R, Onder E, Gunhan O (2006) Malignant fibrous histiocytoma of the mandible. *Dentomaxillofac Radiol* 35:125–128
2. AL-Salihi KA, Samsudin AR (2004) Malignant fibrous histiocytoma (MFH) of the mandible and submandibular salivary gland: report of a case. *Oral Oncol* 40:123–125
3. Sohail D, Kerr R, Simpson RHW, Babajews AV (1995) Malignant fibrous histiocytoma of the mandible: the importance of an accurate histopathological diagnosis. *Br J Oral Maxillofac Surg* 33:166–168

4. Amjad M, Bari AU (2009) Malignant fibrous histiocyoma: an uncommon soft tissue sarcoma. *J Pak Assoc Dermatol* 19:243–246
5. Feldman F, Norman D (1972) Intra and extraosseous malignant histiocyoma (fibrous xanthoma) of bone. *Radiology* 104:497–499
6. Bali A, Singh MP, Padmavathi, Khorate M, Ahmed J (2010) Malignant fibrous histiocyoma: an unusual transformation from benign to malignant. *J Cancer Sci Ther* 2:53–57
7. Vargas A, Echevarria E, Guzman JE (1987) Primary malignant fibrous histiocyoma of the mandible. *J Oral Maxillofac Surg* 45: 634–638
8. Narvaez JA, Muntane A, Narvaez J, Martin F, Monfort JL, Pons LC (1996) Malignant fibrous histiocyoma of the mandible. *Skeletal Radiol* 25:96–99
9. Kanazawa H, Watanabe T, Kasamatsu A (2003) Primary malignant fibrous histiocyoma of mandible: review of the literature and report of a case. *J Oral Maxillofac Surg* 61:1224–1227
10. Abdul-Karim FW, Ayala AG, Chawla SP, Jing BS, Goepfert H (1985) Malignant fibrous histiocyoma of jaws. A clinicopathologic study of 11 cases. *Cancer* 56:1590–1596
11. Anavi Y, Herman G, Graybill S, Macintosh RB (1989) Malignant fibrous histiocyoma of the mandible. *Oral Surg Oral Med Oral Pathol* 68:436–443