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MANAGEMENT OF GIANT CELL MALIGNANT FIBROUS HISTIOCYTOMA (MFH) OF HEAD AND NECK WITH VASCULAR RECONSTRUCTION

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ABSTRACT
Giant cell rich malignant fibrous histiocytoma (MFH) is a mesenchymal tumor and accounts for 3-15% of all cases of MFH. The diagnosis is based on histology and immunohistochemistry that helps in differentiation from other fibrohistiocytic lesions. Giant cell MFH in the head and neck region is a rare entity. We report a case of 67 year old male patient with a left supraclavicular tumor invading the surrounding soft tissue and salivary gland. Radical excision of the tumor and partial excision and patch repair of subclavian vein was done. Histologically, the tumor consisted of spindle shaped cells with prominent mitoses, and abundant, osteoclast-like multinucleated giant cells. Tumor cells showed positive immunoreactivity for Vimentin and Ki-67 while negative for Cytokeratin, SMA, CD34, osteopontin, HMB 45, CD 56, myoglobin and EMA .The giant multinucleated cells were CD68 positive. Therefore the present communication provides an additional case of this rare variant of sarcoma in the head and neck region and its management.

INTRODUCTION

Malignant fibrous histiocytoma (MFH) is a tumor of mesenchymal origin with uncertain histogenesis. This tumor is mainly found in the limbs, abdomen and retro-peritoneum [1]. Histologically there are five subtypes of MFH namely storiform-pleomorphic, myxoid, giant cell, inflammatory and angiomatoid. The giant cell rich malignant fibrous histiocytoma accounts for 3-15% of all cases of MFH [2]. The nomenclature giant cell MFH is currently reserved for 'high grade' undifferentiated pleomorphic sarcomas with prominent osteoclastic giant cells. This entity is considered to be synonymous with malignant giant cell tumor of soft parts [3]. We report a case of giant cell MFH originating as a supraclavicular mass with involvement of the surrounding submandibular gland, sternomastoid muscle, internal jugular vein and subclavian vein. A panel of immunohistochemical stain was performed to establish the diagnosis. It is important to recognize this tumor entity as a distinct giant cell rich variant and differentiate from other giant cell rich pleomorphic sarcomas since therapeutic and prognostic differences are being appreciated currently.

CASE REPORT

We report a case of 67 years old male who presented with left supraclavicular swelling and dyspnoea on exertion for past 6 months. On local examination 8x8 cm hard mass palpable in left supraclavicular fossa, upper limit hyoid bone and lower edge was going till clavicle. Contrast enhanced CT scan (CECT) of neck, chest, abdomen and pelvis was done. CECT neck showed 6x6x7

cm enhancing mass in left side of neck extending 2 cm above hyoid bone till medial end of clavicle with underlying bony erosion over clavicle, however no intrathoracic extension. Mass was displacing left carotid artery and internal jugular vein posteriorly with loss of fat plane with them and infiltrating into submandibular gland, sternomastoid and strap muscles (Figure 1). Mandible, hyoid bone and thyroid cartilage were normal. CECT chest showed two well defined nodules in right upper lobe with normal tracheobronchial tree. Ultrasound guided core biopsy was suggestive of pleomorphic sarcoma but grade of malignancy remained uncertain. Whole body PET scan revealed metabolically active mass in left side of neck (Maximum SUV 33) with metabolically inactive small bilateral lung nodules.

Patient underwent radical excision of mass with clavicle and partial excision with patch reconstruction of left subclavian vein by left supraclavicular approach. Intraoperatively 8x8 cm multilobulated mass was present in left side of neck starting at level of hyoid bone and inferiorly adherent to medial part of clavicle. Mass was infiltrating left sternocleidomastoid muscle, lower half of internal jugular vein and junction of left internal jugular with subclavian vein. Left clavicle and mass was excised en block with left sternomastoid muscle and internal jugular vein. Lower part of internal jugular vein was involved till junction with subclavian vein so partial excision and patch reconstruction of left subclavian vein done (Figure 2). Upper part of internal jugular vein was used for patch reconstruction. Subclavian artery and brachial plexus were preserved and thoracic duct was ligated. Gross examination of the excision specimen shows a tumor measuring 12×9×4 cm with attached part of clavicle. Cut surface of the tumor was homogenous and white with gelatinous area. Microscopic examination showed a malignant mesenchymal tumor composed of pleomorphic ovoid-to-spindle shaped cells arranged in vague nodular pattern and in fascicles. Many multinucleated giant cells were also seen placed among fusiform cell fascicules. Typical and atypical mitotic figures, areas of sclerosis and foci of necrosis were identified. All the resected skin and soft tissue margin and resected clavicle were reported to be free of tumor infiltration. A panel of immunohistochemistry was performed using pancytokeratin, vimentin, smooth muscle actin, CD68, CD34, osteopontin, S100, HMB 45, CD 56, CD68, myoglobin, EMA and Ki67. Vimentin was diffusely positive in the tumor cells. The histiocytic marker CD68 was positive in large and giant tumoral cells. S100 was variable positive in spindle-shaped cells (Figure 3). Smooth muscle actin was positive in vessels within the tumor mass. CD34 was positive in vessels and negative in tumor. The Ki-67 labelling index was approximately 40-50% in the highest proliferative area. The tumor cells were immunonegative for pancytokeratin, EMA, myoglobin, osteopontin, HMB45 and CD 56. Special histochemical stain VVG did not reveal any osteoid production. Based on above histological and immunohistochemical features, a diagnosis of Giant cell MFH was made.

Postoperative recovery was uneventful and patient received adjuvant chemotherapy and radiotherapy. On follow up after 6 months no local recurrence or distant metastasis observed.

DISCUSSION

The term MFH was introduced by Kauffman and Stout in 1961 to describe a histiocytic-like tumor with predominant fibroblast [4]. Approximately 3 to 10% of all MFHs are localized in the head and neck and the most common localizations are in the sinonasal cavities (30%), the cranio-facial bones (15–25%), larynx (10–15%) and soft tissues of neck (10-15%) [1, 2].

Age is an important determinant of histological type of soft tissue sarcoma (STS). Most MFH patients are above 40 years at presentation. 80% cases of head and neck STS present with painless neck mass. Visual disturbance, epistaxis, chronic sinusitis, otolagia, sensory and/or motor disturbances are the other presenting features [5]. Imaging findings are nonspecific, however computed tomography scan (CT scan) and Magnetic resonance imaging (MRI) are essential for preoperative staging, loco regional assessment and surgical planning and preferably to be done before biopsy [6].

Current literature classifies MFH as ‘high grade’ undifferentiated pleomorphic sarcoma (HGUPS). Contrary to the previous data, the 2002 WHO classification estimates MFH to be less than 5 % of all adult sarcomas [3, 7]. Histologically there are five subtypes of MFH that includes pleomorphic, myxoid, inflammatory, giant cells and angiomatous variant. The giant cell variant accounts for less than 15% of all MFH [7]. No specific immunohistochemical marker exists for MFH, but immunohistochemistry (IHC) can be used to differentiate other malignancies from MFH as most often it remains a diagnosis of exclusion. The histological examination corroborates IHC in making a definitive diagnosis [3, 8]. In their classic description, Weiss and Enzinger [9] described MFH to be composed of a combination of ‘spindle cell (fibroblast-like) and histiocyte-like cells arranged in a storiform pattern and accompanied by pleomorphic giant cells and inflammatory cells’. In the presented case, histological study confirmed giant cell MFH and immunohistochemistry confirmed the mesenchymal origin of the tumor (vimentin and CD68-positivity), allowed differentiation from a sarcomatoid carcinoma (pancytokeratin, EMA negativity), melanoma (HMB45-negativity), angiosarcoma (CD34-negativity), leiomyosarcoma (absent fascicular pattern/ smooth muscle morphology and SMA –negativity) and rhabdomyosarcoma (absent alveolar/ striated muscle structure and myogenin-negativity); Ki67-positivity in 40–50% of tumor cells confirms malignancy and proves aggressiveness of the tumor. In our case, the tumor cells were focally immunoreactive for S100 protein pointing

towards neurogenic tissue but neurogenic sarcoma was ruled out due to histological characteristics. Hence MFH could be differentiated from carcinomas, rhabdomyosarcomas, leiomyosarcomas and similar malignancies based on a combination of histology and IHC pattern.

Wide local excision with negative margins is treatment of choice but not feasible in most head and neck sarcomas due to complex anatomy and proximity with major vital structures. Every attempt should be made to achieve negative margins without cosmetic and functional compromise [5, 6]. Soft tissues tumors of supraclavicular region have propensity to invade neural and vascular elements [10]. Vascular infiltration is usually limited to adventitial layer allowing subadventitial dissection without resection [11]. When subclavian vein resection is necessary for oncological reasons, after removal of involved part simple ligation is preferred approach due to rich collateral circulation [10]. However to avoid venous hypertension, revascularization should be considered in case of poor venous collateralization [12]. Although short defects can be repaired directly without luminal compromise, bypass with interposition of an autologous vein or polytetrafluroethylene graft remains standard approach [11]. After oncological resection with lymphadenectomy and possible adjuvant radiotherapy the risk of infection becomes significant and venous material may be preferred in such cases [12]. In the present case, the tumor was infiltrating Left IJV and subclavian vein junction so after en bloc excision of tumor with IJV venous patch was taken from upper part and used to repair the defect of subclavian vein. Although this technique has not been reported in literature it has many advantages: relatively simple procedure in which only one wall of subclavian vein needs to be repaired, less chances of luminal compromise, no need for prosthetic material and less incidence of infection. Tumour grade, size, depth, histological subtype, distant metastases at initial presentation and ability to achieve local control seem to be the important prognostic factors for MFH [5, 6, 13]. Head and neck MFH has poorer prognosis, compared with MFH elsewhere in the body. Recently, Sabesan et al reported a 48% 5-year survival rate for MFH of the head and neck and 77% for MFH of the trunk and extremities. Reported rates of local recurrences (86% after marginal excision, 66% after wide excision and 27% after radical resection) highlighted the importance of radical resection [14]. Adjuvant chemotherapy and radiotherapy seem to be critical for optimal treatment outcome [5, 6].

Fig 1. CECT Neck of the patient showing large heterogeneous mass on the left side which is compressing the left internal jugular vein and displacing the trachea towards right side.

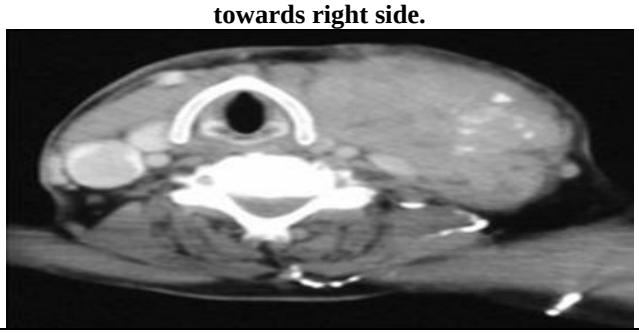
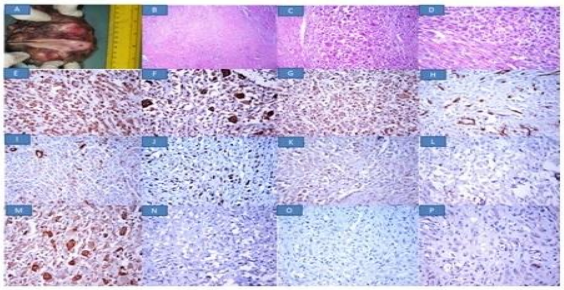


Fig 2. Intra-operative photograph after tumor resection showing patch repair of left subclavian vein

Fig 3. (A) Gross photograph showing a tumor(12×9×4)cm with homogeneous grey-white cut surface.(B) Low power photomicrograph showing a malignant mesenchymal tumor composed of oval to spindle shaped cells with areas of sclerosis (x20; H&E). (C), (D) Many multinucleated giant cells are seen placed among fusiform cell fascicules (X20; H&E). (E) On immunohistochemistry tumor cells are immunopostitive for Vimentin (X40), (F) CD68 (X40), (G) Variable S-100 positivity in spindle shaped cells (X40) and are (H) immunonegative for CD34 (X40), (I) SMA (X40), (K) Pancytokeratin (X40), (L) EMA (X40), (M) Osteopontin (X40), (N) Myogenin (X40), (O) HMB-45(X40) and (P) Chromogranin (X40). (J) KI-67 labeling index is high (approx. 40-50%) (X40).



CONCLUSION

Surgical resection remains primary therapy in MFH of head and neck region. Because of the rarity of tumor large scale prospective randomized trial might not be feasible as far as adjuvant treatment is concerned. As giant cell variant of MFH has aggressive behavior and poor prognosis, adjuvant radiotherapy with or without chemotherapy should be considered based on retrospective data.

CONFLICT OF INTEREST

The author(s) declare that they have no conflict of interest.

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STATEMENT OF HUMAN AND ANIMAL RIGHTS

All procedures performed in human participants were in accordance with the ethical standards of the institutional research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards. This article does not contain any studies with animals performed by any of the authors.

REFERENCES

1. Kasat VO, Saluja H, Rudagi BM, Kalburge JV and Sachdeva S. (2014). Malignant fibrous histiocytoma of maxillary alveolar ridge extending to the hard palate. *J Cancer Res Ther,* 10, 422-424.
2. Anghelina F, Ionita E, Chiutu L, Mogoanta CA, Ciolofan S, Iosif C and Ceausu M. (2009). Malignant fibrous histiocytoma of larynx with giant cell: case report and histological-clinical considerations. *Rom J Morphol Embryol,* 50, 481-5.
3. Fletcher CD. (2006). The evolving classification of soft tissue tumors: an update based on the new WHO classification. *Histopathology,* 48, 3–12.
4. Kauffman SL and Stout AP. (1961). Histiocytic tumors (fibromas xanthoma and histiocytoma) in children. *Cancer,* 14, 469–82.
5. Patel SG, Shaha AR and Shah JP. (2001). Soft tissue sarcoma of the head and neck: an update. *Am J Otolaryngol,* 22, 2–18.
6. Aljabab AS, Nason RW, Kaz Ri and Pathak KA. (2011). Head and Neck Soft Tissue Sarcoma. *Indian J Surg Oncol,* 2, 286–290.
7. Matushansky I, Charytonowicz E, Mills J, Siddiqi S, Hricik T and Cordon-Cardo C. (2009). MFH classification: differentiating undifferentiated pleomorphic sarcoma in the 21st Century. *Expert Rev Anticancer Ther,* 9**,** 1135–1144.
8. Qiu SQ, Wei XL, Huang WH, Wu MY, Qin YS, Li YK and Zhang GJ. (2013). Diagnostic and therapeutic strategy and the most efficient prognostic factors of breast malignant fibrous histiocytoma. *Sci Rep,* 3, 25-29.
9. Weiss SW and Enzinger FM. (1978). Malignant fibrous histiocytoma. An Analysis of 200 Cases. *Cancer,* 41, 2250–66.
10. Dartevelle PG, Chapelier AR, Macchiarini P, Lenot B, Cerrina J, Ladurie FL, Parquin FJ and Lafont D. (1993). Anterior transcervical–thoracic approach for radical resection of lung tumors invading thoracic inlet. *J Thorac Cardiovasc Surg,* 105, 1025-34.
11. Fadel E, Chapelier A, Bacha E, Leroy-Ladurie F, Cerrina J, Macchiarini P and Dartevelle P. (1999). Subclavian artery resection and reconstruction for thoracic inlet cancers. *J Vasc Surg,* 29, 581-8.
12. Gonzalez M, De´glise S, Ris HB and Corpataux JM. (2010). Reconstruction of a resected subclavian vein by transposition of the ipsilateral internal jugular vein. *J Thorac Cardiovasc Surg,* 140, 1198-9.
13. Weiss SW and Goldblum JR. (2008). Malignant fibrous histiocytoma (pleomorphic undifferentiated sarcoma). In: Weiss SW, Goldblum JR: Enzinger and Weiss's Soft Tissue Tumors. 5th ed. St. Louis, Mo: Mosby, 403-27.
14. Sabesan T, Xuexi W, Yongfa Q, Pingzhang T and Ilankovan V.(2006). Malignant fibrous histiocytoma: outcome of tumors in the head and neck compared with those in the trunk and extremities. *Br J Oral Maxillofac Surg 4*4, 209–12.