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Case Report

Maxillary aggressive angiomyxoma showing ineffective to radiotherapy: a rare case report and review of literature

Zhaoling Wang*, Ying Liu*, Lu Yang*, Ling Gu, Yuhong He, Diyan Huang, Guoxiong Zhu

Department of Stomatology, General Hospital of Jinan Military Command, Jinan 250031, Shandong, China.
**Equal contributors.*

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Abstract: Aggressive angiomyxoma, mostly originating in the female pelvis and peritoneum or in the male analogous sites, is a rare mesenchymal neoplasm characterized with infiltrative growth to adjacent tissue and local recurrence after primary excision. Herein, we report a case of aggressive angiomyxoma of maxilla in a 60-year-old male patient for its rarity. The patient presented with a one-year history of progressively enlarging maxillary mass on left side. Before referred to our hospital, he was given a biopsy and diagnosed as aggressive angiomyxoma by immunohistochemical staining. After that, he underwent 60Gy radiotherapy. Unfortunately, CT scan showed bigger mass infiltrated to adjacent facial soft tissues and bones compared with that of before radiotherapy. Besides that, he began to suffer with ingravescent headache. The mass was surgically removed and the diagnosis was confirmed by immunohistology in our hospital. As a case of aggressive angiomyxoma occurred in a rare site and experienced an ongoing growth in spite of radiotherapy, its characteristics was discussed with a brief literature review, which may aid further understanding of aggressive angiomyoma.

Keywords: Maxillary neoplasm, aggressive angiomyoma, radiotherapy, treatment

Introduction

Aggressive angiomyxoma, first described by Steeper and Rosa in 1983 [1], is a rare mesenchymal neoplasm and the term "aggressive" means it is characterized by a locally aggressive growth pattern and has a potential of local recurrence rather than metastatic potential [2]. Aggressive angiomyxoma occurs predominantly in the pelvic and perineal regions in adult females of reproductive age and arises sometime in the analogous sites of older males [3]. Only a very few cases were reported involving extragenital sites [4-9]. Here, we report a case of aggressive angiomyxoma of maxilla in a 60-year-old male patient for its rarity. To the best of our knowledge, this is the first case of maxillary aggressive angiomyxoma in English literature.

Case report

A 60-years-old male was referred to our hospital with a one-year history of progressively enlarging maxillary mass and ingravescent headache on left side. At first he sought treatment in a local clinic with complaint of a mass like a peanut on his left posterior maxillary. He was thought suffering with soft tissue infection and given some prescription. The treatment seemed ineffective to the mass and a new complaint of headache appeared. The patient then went to a hospital where he was found an ill-defined low density mass of left maxillary by CT scan (**Figure 1A**). Then, he underwent a biopsy and diagnosed as aggressive angiomyxoma. There he was given a 60-Gy irradiation. Unfortunately, the irradiation therapy seemed ineffective because the mass was found obviously growing bigger by CT scan 3 months later (**Figure 1B**). The patient was suggested to our hospital for surgery. On physical examination a swelling of left maxillary was confirmed which had extend to the right palate. Teeth from #8 to #15 were seriously loose. The tumor was surgically removed by relatively complete excision and no signs of regional recurrence or distant metastasis were observed during the follow-up period of 2 months.

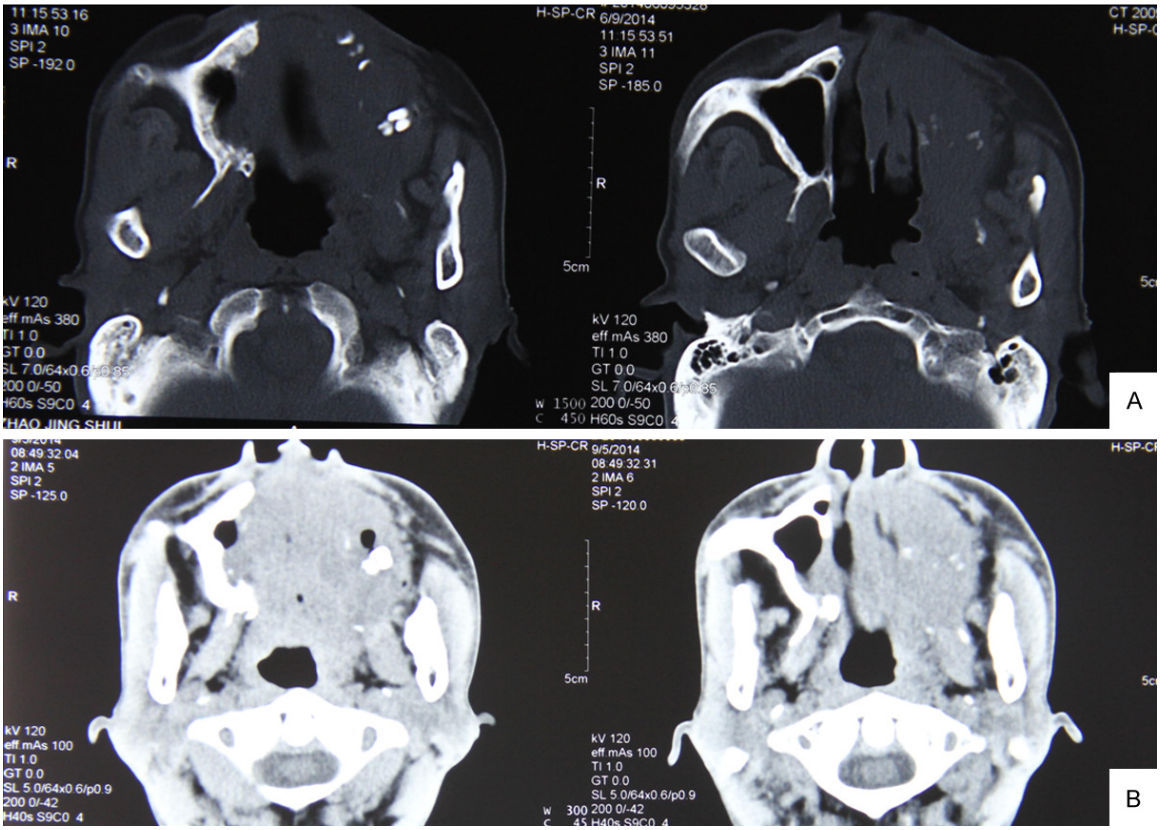

Figure 1. CT scans showing: A. maxillary hypodense mass with obscure border. B. increased mass volume with extensive infiltration to adjacent tissues after radiotherapy. Note the involved nasal septum and the lost tissue spaces.

Surgically, this tumor was unencapsulated and characterized by infiltration to the adjacent organs. Except that the left maxilla and palate was entirely destroyed, left zygoma, left sphenoid, right maxilla and palate was partly destroyed as well as the adjacent soft tissues. Excision of this tumor with a 1.0-cm clear margin was made except the left infraorbital region for the sake of preservation of the function of the left eye. Macroscopically, the tumor showed moderate hardness and a grayish white cut surface with obscure boundary. Histological examination of the tumor revealed scattered spindle and satellite shaped fibroblastic cells set in myxoid matrix with prominent vascularity (**Figure 2A**). The tumor was poorly circumscribed and exhibited infiltration into adjacent tissues such as muscles, salivary glands and bones. Immunohistochemically, the tumor cells were positive for vimentin (**Figure 2B**), smooth muscle actin (**Figure 2C**), CD34 and Ki-67 (10%), and a few of cells were positive for estrogen receptor, but negative for S-100 (**Figure 2D**), progesterone receptor and desmin.

Discussion

As a rare and locally aggressive myxoid mesenchymal neoplasm, aggressive angiomyxoma occurs predominantly in women, with a female to male ratio of approximately 6:1 [10, 11] and is mostly reported as cases reports and small series. Because of its rarity and non-specific symptoms, early diagnosis is difficult and usually made by histopathology. In literatures, immunoreactivity of vimentin, desmin, smooth muscle actin, muscle-specific actin, CD 34, estrogen and progesterone receptors is more often used for diagnosis and differential diagnosis of aggressive angiomyoma. Generally the tumor shows immunopositivity for vimentin, desmin, smooth muscle actin and immumonegativity for S100, but immunopositivity for estrogen and progesterone receptors varies between females and males [3, 11-13]. In this male case, there were a few of cells positive for estrogen receptor and whether it has clinical meaning should be further studied. However, there is no specific immunohistochemical marker for aggressive angiomyoma.

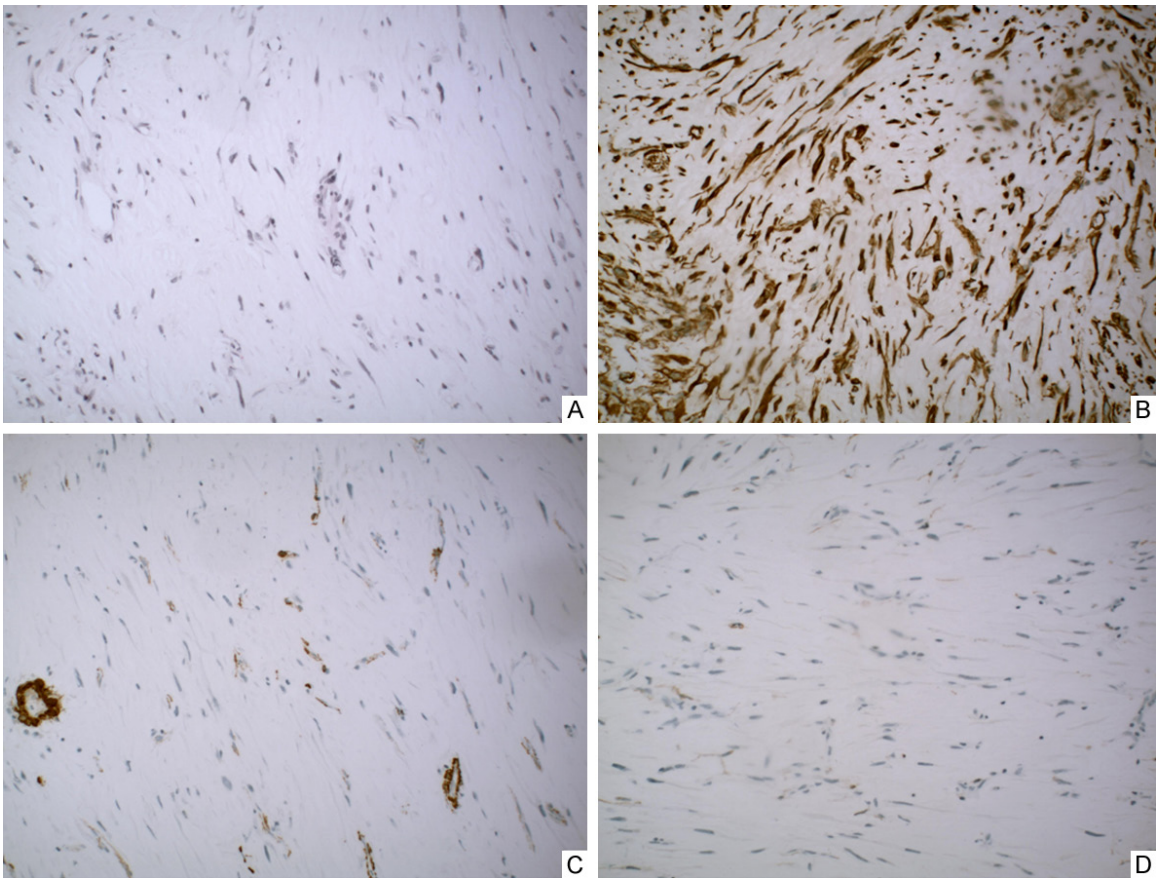

Figure 2. Microscopic findings and immunohistochemical staining. A. Histological examinations stained with hematoxylin and eosin revealed scattered spindle and satellite shaped fibroblastic cells set in myxoid matrix with prominent vascularity, × 200. B. Immunohistochemical staining showed that tumor cells was positive for vimentin, × 200. C. Immunohistochemical staining showed that tumor cells was positive for smooth muscle actin, × 200. D. Immunohistochemical staining showed that tumor cells was negative for S-100, × 200. Preoperative clinical view of lesion of the oral floor and sublingual region.

Since pre-surgery diagnosis is difficult, ultrasound imaging, CT scan and MRI are usually used to reveal the extension of this tumor and helpful for surgery design [2, 3, 14]. Unlike tumors occurred in soft tissues, this rare maxillary case extensively involved the adjacent soft tissues and facial bones which cause the symptom of loose teeth and headache. CT scan illustrated an appearance of great destructivity like that of maxillary malignant tumors. It indicated that as a mass in bones, CT scan may be more helpful to decide the infiltration and thus guide the surgery than MRI and ultrasound imaging. Considering that there are many sinus and cavities in maxillofacial region and the tissues are relatively loose, the extensive infiltration of this maxillary aggressive angiomyxoma may due to anatomical peculiarities of this region.

In spite of reported adjuvant therapy of radiotherapy, chemotherapy, vessel embolization, and hormone suppression, there are still not many management options available for aggressive angiomyxoma, and a radical surgical resection is still the first line of treatment [1, 3, 10-14]. Although radical excision seemed to bring down local recurrence which is the significant cause of patient morbidity, a review of over 106 cases questioned this belief and showed that there is no significant difference of recurrence between patients with clear resection margins and those having tumor-involved resection margins [2, 11]. Nevertheless, achieving negative resection margins is not always easy because of the tuomor's infiltration and undefined capsule. For cases with extensive infiltration or occurring in deep-seated organs, rather than smaller or superficial cases, radical excision may confer a higher danger of severe surgical morbidity such as excessive blood loss, associated organ dysfunction. However, aggressive angiomyxoma is

generally not life-threatening and the prognosis for patients is good except two exceedingly rare cases of metastases. In light of these facts, radical excision may not be justified and an incomplete removal is acceptable with long term follow-up [1, 3, 11, 15]. As for our case, infraorbital tissues were infiltrated and more radical excision would sacrifice the left eye. In addition, as tumors of maxillofacial region, there is much limitation for radical excision because of the vital organs nearby. Weighing the advantages and disadvantages, a relatively complete excision was chosen for this case and the left eye was preserved. With a 2-month follow-up, no hints of recurrence were found, and also, a continuing follow-up are emphasized to determine the long-term prognosis of this very rare maxillary aggressive angiomyxoma.

Although radiotherapy was reported as an adjuvant therapy for some recurrent cases and did control the tumor [16], in this case, aggressive angiomyxoma kept growing under irradiation therapy, which indicated the limitation of radiotherapy for this tumor, at least to this case. Or, in the other hand, there may be of some biological behavior difference between maxillary aggressive angiomyxoma and those occurred in soft tissues, or pelvic region and analogous region.

In summary, we here reported a very rare case of maxillary aggressive angiomyxoma which underwent radiotherapy first but seemed no effect. Long-term follow-up and further cases would be helpful to determine whether maxillary angiomyxoma has the same biological behaviors as its pelvic counterpart. This is an interesting case and may aid further understanding for aggressive angiomyxoma, especially extragenital cases.

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Disclosure of conflict of interest

None.

Address correspondence to: Dr. Guoxiong Zhu or Dr. Yuhong He, Department of Stomatology, General Hospital of Jinan Military Command, Jinan 250031, Shandong, China. Tel: +86 531 51665976; +86 531 51666077; E-mail: jzkqk@sina.com (GXZ); callushong@163.com (YHH)

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