

Case Report

Hemangiopericytoma associated with multiple keratocystic odontogenic tumors in an adolescent patient: a case report

Rajdeep Brar¹⁾, Sunita Kulkarni¹⁾, Soheyl Sheikh¹⁾, Sanjeev Jindal¹⁾ and Prabhleen Brar²⁾

¹⁾Department of Oral Medicine and Radiology, M.M College of Dental Sciences and Research, Ambala, India

²⁾Department of Conservative Dentistry, M.M College of Dental Sciences and Research, Ambala, India

(Received 3 December 2007 and accepted 1 April 2008)

Abstract: Hemangiopericytoma, initially described by Stout and Murray in 1942 (1), is a rare vascular tumor arising from mesenchymal cells with pericytic differentiation. Hemangiopericytomas usually occur in the 5th decade of life and account for 3-5% of all soft tissue sarcomas and 1% of all vascular tumors (2). The tumor usually occurs in the limbs, pelvis, or head and neck region; 15-30% of all hemangiopericytomas occur in the head and neck (2,3). Here we present a case of hemangiopericytoma of the submandibular region with keratocystic odontogenic tumors in an adolescent patient. (J. Oral Sci. 50, 233-237, 2008)

Keywords: hemangiopericytoma; odontogenic keratocyst; immunohistochemistry; adolescent.

Introduction

Hemangiopericytoma is a rare vascular tumor comprising 1% of all vascular neoplasms in adult life (2), and one third of these lesions occur in the head and neck region (4). Hemangiopericytomas are thought to arise from mesenchymal cells with pericytic differentiation.

Pericytes line the epithelial cells in the capillary walls. They were originally described by Rouget in 1873 as pericapillary ameboid cells, and he termed them cellular adventicities (5). In 1923 Zimmermann named these cells pericytes (5). Pericytes are mesenchymal in origin and are

relatively undifferentiated in the sense that they can apparently develop into several different cell types, including smooth muscle. They are contractile and can change their shape, thereby reducing the diameter of the capillary lumen (6).

In 1942 Stout and Murray were the first to describe tumors of pericytes and they coined the term hemangiopericytoma (1). In the following 60 years, the term has been loosely used to describe a wide range of neoplasms with certain morphological characteristics such as: a monotonous appearance on low-power examination, moderate to high cellularity, and the presence of numerous, branching 'staghorn vessels' with walls of variable thickness (7).

Hemangiopericytoma cells usually have uniform, ovoid nuclei and indistinct cell borders. They are located outside the reticulin sheath of the epithelium.

We report a case of hemangiopericytoma of the submandibular region with multiple keratocystic odontogenic tumors in the maxilla and mandible in a 15-year-old female patient.

Case Report

A 15-year-old girl consulted the department of Oral Medicine and Radiology with a chief complaint of swelling in the left sub-mandibular region of two months' duration. The swelling was initially very small and gradually increased to the present size (Figs. 1 and 2).

The patient revealed that she had developed a swelling in the same region 2 years earlier; this was diagnosed as myxoid tumor and was subsequently removed surgically. One year later she was diagnosed with multiple keratocystic odontogenic tumors in the maxilla and mandible. The mandibular and left maxillary lesions were excised, sent for histopathological examination, and confirmed as

Correspondence to Dr. Rajdeep Brar, Department of Oral Medicine and Radiology, MM College of Dental Sciences and Research, Mullana, Ambala, India

Tel: +91-1722580512

Fax: +91-1731274777

E-mail: drrajdeepbrar@yahoo.co.in

keratocystic odontogenic tumors. When she came to our department she brought her previous panoramic radiograph taken one year previously. This showed a well-defined radiolucency in the mandible extending from the mesial aspect of 46 and crossing the midline to the mesial aspect of 33. A second radiolucency was seen from the distal aspect of 37 extending posteriorly to the ramus and inferiorly to the lower border of the mandible. A third radiolucency was seen in the left maxillary region extending from the distal aspect of 22 to the mesial aspect of 24, associated with an impacted 23 and an overretained 63.



Fig. 1 Extra-oral photograph of the patient showing the swelling in the submandibular region.



Fig. 2 Lateral view of the lesion.

On examination the present swelling on the left side of the neck and submandibular region measured 5×3 cm. The swelling extended anteriorly to the symphysis, posteriorly to the angle of the mandible, superiorly to the base of the mandible, and inferiorly to the level of the thyroid cartilage.

The skin over the swelling was stretched, normal in color, and pinchable. The swelling was non-tender, non-pulsatile, non-fluctuant, movable, and not fixed to the underlying structures. There was no lymphadenopathy.

Based on the clinical findings, we arrived at a provisional diagnosis of rhabdomyosarcoma with a differential diagnosis of pleomorphic adenoma of the submandibular gland.

The patient then underwent various investigations.

Radiographic findings

Panoramic radiography showed a well-defined radiolucent lesion in the right maxillary region extending from the distal aspect of 12 to the mesial aspect of 16 with a well-defined radiopaque margin suggestive of an odontogenic keratocyst (Figs. 3, 4). It is interesting to

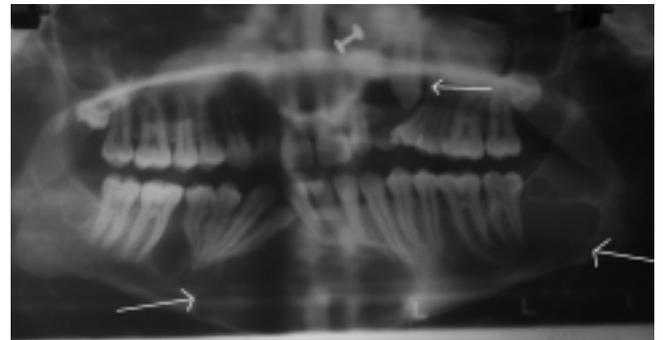


Fig. 3 Panoramic radiograph showing multiple odontogenic keratocysts, one year before surgery of hemangiopericytoma.

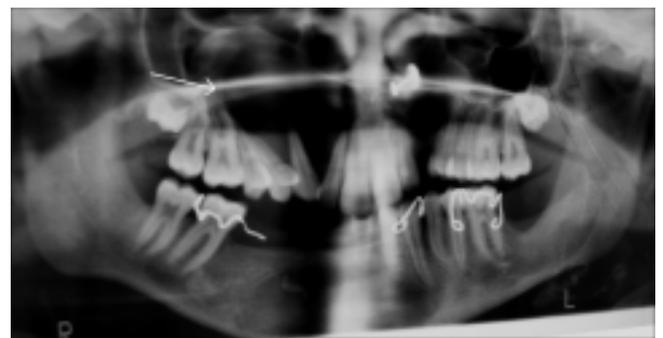


Fig. 4 Panoramic radiograph before surgery, showing single odontogenic keratocyst in the right maxilla.

note that this was not present in the panoramic radiograph taken a year earlier.

Other radiolucencies seen on this radiograph in the mandibular midline, left mandibular region, and left maxillary region correlated with the previous history of treatment of multiple keratocystic odontogenic tumors.

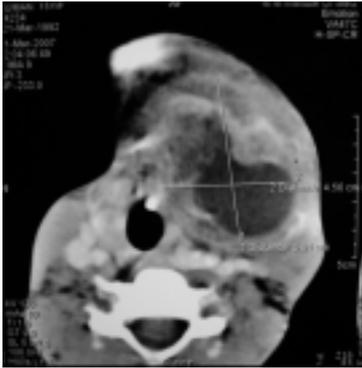


Fig. 5 Measurements of the lesion on axial view.

Computed tomography findings

Computed tomography revealed a heterogeneous predominantly hypodense mass of approximately 6 × 5 × 6 cm in the left submandibular and neck area (Fig. 5). On computer-enhanced computed tomography, central necrosis was visible in the large soft tissue mass and a peripheral thick soft-tissue component showed enhancement. Anteriorly there was no peripheral calcification in the mass (Figs. 6-9).

Superiorly the mass extended from the left submandibular area, and inferiorly it was hanging exophytically to the level of the lower border of the thyroid. Medially it extended to the paralaryngeal area with compression of the larynx. However, the cartilages of the larynx, the hyoid bone, and the thyroid gland were normal. Laterally it extended into the soft tissue causing compression and displacement of the neck vessels; however, no involvement, invasion, or thrombosis of these vessels was seen.

A well-defined intra-osseous cyst was noted in the right side of the maxilla, arising from the alveolar margin. No bone fenestration was present but the cyst extended into

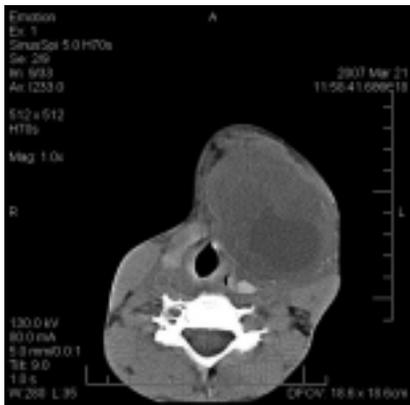


Fig. 6 CT scan showing axial view of the lesion.



Fig. 8 CT scan showing coronal view of the lesion.

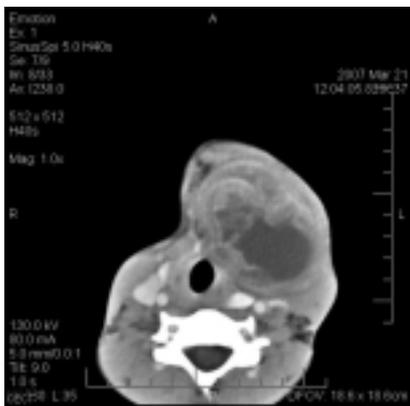


Fig. 7 CT scan showing axial view of the lesion.



Fig. 9 CT scan showing coronal view of the lesion.

the maxillary sinus.

Histopathologic findings

Excisional biopsy of the lesion was performed and the tissue sent for histopathological examination.

Sections stained with hematoxylin and eosin showed a highly cellular tumor with tumor cells ranging from round to oval to spindle shaped, with round to oval nuclei seen proliferating around the blood vessels. Tumor cells were

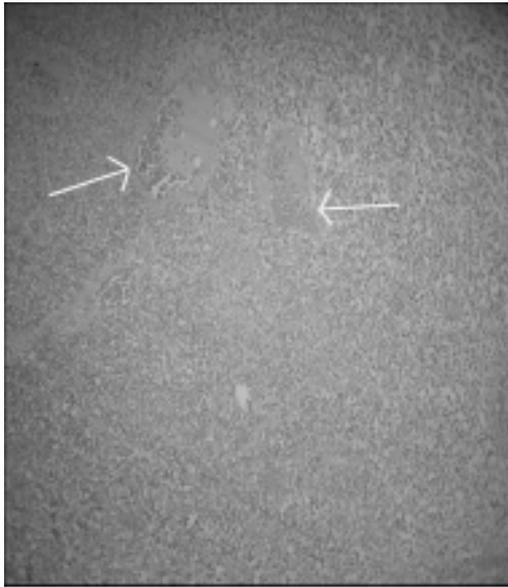


Fig. 10 Histopathological slide of the lesion showing the staghorn pattern of vessels (arrow, $\times 100$ magnification).

arranged in sheets. Few mitotic figures were seen under high power. The connective tissue stroma was scanty, and few myeloid areas were seen. The section also showed numerous blood vessels of varying caliber lined by endothelium. Some blood vessels were dilated and some exhibited a 'staghorn pattern' of branching, suggestive of hemangiopericytoma (Fig. 10).

Histopathological examination of the keratocystic odontogenic tumor was shown in Fig. 11.

After routine histopathological examination the specimen was sent for immunohistochemistry. The tumor cells were positive for vimentin and negative for S100 protein, smooth muscle actin, desmin, CD31, and CD34. (Fig. 12)

Discussion

This case deserves special attention because of the many unusual features. This tumor mostly arises in the 4-6th decade of life and is rarely seen in the 2nd decade, as in the present case. Hemangiopericytoma is commonly seen in the pelvis and limbs, and only 15-30% of all vascular neoplasms occur in the head and neck region (1,2,8) of which most are seen in the paranasal sinuses, tongue, and palate (9). To the best of our knowledge, this is the first case of hemangiopericytoma in the submandibular region. Another very unusual feature, and one that we do not believe has been reported previously, is the concurrent presence of multiple keratocystic odontogenic tumors in the mandible and maxilla.

Hemangiopericytoma usually occurs deep in the soft tissue, grows insidiously, and may be associated with pain. Clinically this tumor has a grey-white or brownish

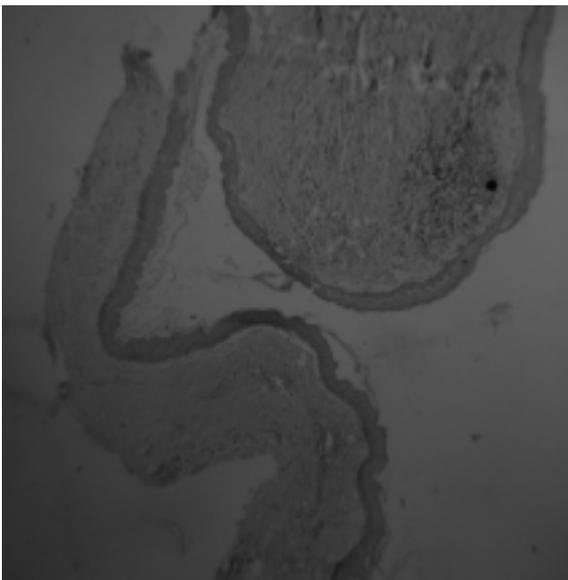


Fig. 11 Histopathological photograph of the cystic lesion ($\times 100$ magnification).

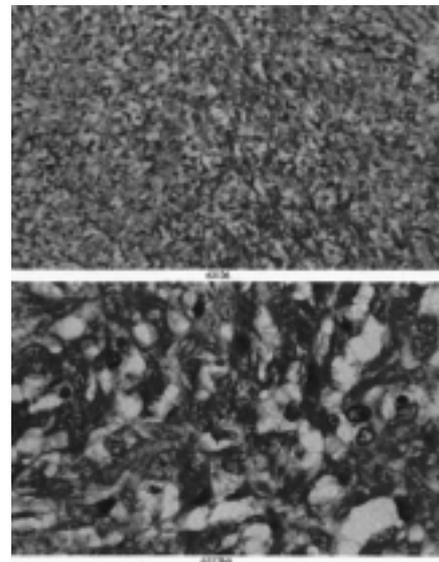


Fig. 12 Immunostaining for vimentin ($\times 400$).

cut surface, and size may vary from 1-20 cm (8). Macroscopically hemangiopericytoma has a well-defined capsule. It can be lobulated or nodular; firmly attached to the muscle or fascia; and soft, spongy, firm, or friable. Microscopically it typically consists of small, closely packed cells with ill defined cytoplasm and darkly stained nuclei. Interspersed between cells are many vascular spaces which may be slit like or sinusoidal. Occasionally the pericytes may have a palisaded configuration or may show interstitial mucoid degeneration (10). Radiographically the tumor consists of a well-circumscribed, radiopaque soft tissue mass that often displaces neighboring structures (11).

It is significant to note that vimentin is the only marker that is consistently expressed in hemangiopericytoma (4).

The management of hemangiopericytoma involves wide surgical excision. In the head and neck, cervical lymphadenectomy is reserved for those instances where palpable adenopathy is coexistent. The role of radiotherapy has been questioned because these tumors are generally radioresistant; one study showed that only 13% of patients were cured with radiotherapy (9).

The present patient underwent wide excision. Radiotherapy was not advised as there was no lymphadenopathy. The patient has been kept under observation and regular follow-up, and remains free of recurrence at the time of writing, 12 months after operation.

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