

# Paediatric Angiosarcoma of Gingivobuccal Sulcus: A Rare Case Report

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## ABSTRACT

We are presenting a case of 10 years male child with huge left sided facial swelling with intermittent bleeding from oral cavity. He underwent total maxillectomy and wide local excision. Angiosarcoma is a rare aggressive malignant case of the pediatric population which occur in head and neck region<sup>1-8</sup>.

**KEY WORDS:** Angiosarcoma, Head and Neck Malignancy, Pediatrics.

## INTRODUCTION

Angiosarcomas are malignant mesenchymal tumors with a differentiation into vascular endothelium. They are rare malignant tumors, predominantly affect adults and elderly patients and are even rarer in pediatric population- a review of 20 years shows a total of 12 cases reported. These locally destructive aggressive high grade tumours with metastatic potential are more aggressive in children<sup>1</sup>. When occurring in children they have predilection to male children and head and neck region. They usually come with clinical presentation of an enlarging, rapidly progressive mass with occasional acute hemorrhage, anemia, or coagulopathy<sup>1,3</sup>. The etiology of angiosarcoma is not fully understood. Sun exposure has been implicated in the etiology of this tumor and the overall prognosis of angiosarcoma is poor even compared with that of other head and neck sarcomas<sup>8</sup>. It is highly aggressive and can arise in association with chronic lymphedema and irradiation. The most common sites are soft tissue and skin. It represents less than 1% of all malignant tumors<sup>12</sup>. Angiosarcoma of the oral cavity is extremely rare; there are a few case reports and case series in the literature. Metastatic spread is usually

hematogenous and a common site of involvement is the lungs. Angiosarcoma of the oral cavity may occur in various tissues such as oral soft tissue, minor salivary glands and bones<sup>13</sup>.

## CASE REPORT

A 10 year old boy presented with swelling of the left cheek along with intermittent bleeding from the oral cavity for four months (Fig.1). There was a small swelling, initially appeared in the left sided upper gingivo-buccal sulcus with mild swelling of left cheek. It used to bleed on brushing teeth but the size was rapidly increasing within last one week and was associated with dull pain, facial swelling, loosening of teeth and spontaneous bleeding from the mouth. The patient was unable to open his mouth and couldn't chew and swallow properly for last one week. There was no history of nasal, aural and eye symptoms. His family history was not significant. There was no history of exposure to radiation. His family was farmer by profession and his younger sibling had no such complaints. He underwent an incisional biopsy under local anesthesia from the oral cavity mass two months back in other hospital. It was reported as Epithelioid hemangioma.

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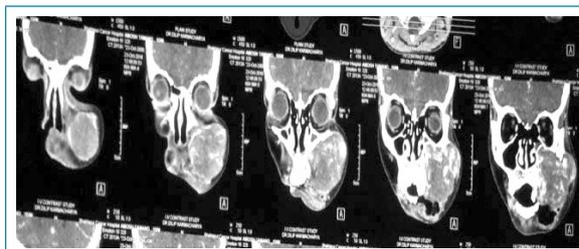
**Fig. 1 Swelling on face.**

On clinical examination, he had huge mass (Fig.1), about 7 x7cms covering the whole of his left side of the face with prominent vessels on the overlying skin, also there was an ulcerative bleeding mass occupying the left upper gingivo-buccal sulcus (Fig.2) that had eroded and deformed the left maxillary alveolar process and teeth. It had extended externally into the cheek - a smooth, fixed, tensed, mild tender mass. His general condition was poor with anemia and dehydration. Examination of nose and ear was clinically normal. His gross vision and eye movements were normal. There was no cervical lymphadenopathy on clinical examination.



**Fig.2 Gingivolabial mass.**

The boy was admitted in the ward with a differential diagnosis of left gingivobuccal Epithelioidhemangioma. Dehydration was corrected, also anemia (Hb-7.2 gm%) was corrected by transfusing two units of packed cell blood. Feeding was managed by placing nasogastric tube. Imaging (CECT) suggested of a locally advanced tumour arising from left hard palate and comprising some vascular component that had eroded left maxilla's lateral and posterior walls and extended into the infratemporal fossa (Fig.3). However overlying facial skin, subcutaneous tissue, nasal cavity, orbit, base of skull and mandible left unaffected.



**Fig. 3 CECT finding.**

We consulted the case with radiologist, dental surgeon and oncologist. Then we planned for wide excision of mass through transzygomatic approach and total maxillectomy. Airway was secured by tracheostomy under local anesthesia pre-operatively. Then general anesthesia was given and excision of mass in total with total maxillectomy and placement of temporary palate prosthesis was done. The intra-operative findings was a highly vascular well circumscribed huge (11x10cm) soft tissue mass (Fig.4) eroding the hard palate, lateral wall and posterior wall of left maxillae. Postoperative period was uneventful and histopathology report confirmed it as well differentiated angiosarcoma. Histology sections was composed proliferating vascular channels and papillary structures lined by atypical endothelial cells with moderate amount of cytoplasm and round to oval vesicular nuclei and prominent nucleoli, moderate cellular pleomorphism large area of haemorrhage and necrosis were present as shown in Fig.5. Immunohistochemistry was advised but patient refused. Case was discharged after closer of tracheostomy stoma with satisfactory cosmetic and general condition from our side. He was referred to radiotherapy department for radiotherapy and advised follow up after radiotherapy. But the patient lost for follow up and he came to our OPD after 9 months. During follow up, his general condition was good and on examination of oral cavity there was mild

trismus with no obvious mass or lesion in the primary site. He had completed his radiotherapy seven months back according to radiotherapy document. .

## DISCUSSION

There is inconsistency in the data on age group involvement of this tumor because some authors postulate that older patients have greater propensity while others suggests that it is more in younger age<sup>14</sup>. Our case was a 10 year old male child. Fanburg-Smith et al suggest a mild male predominance (ratio 1.07:1)<sup>9</sup>. The median age of those who develop angiosarcoma, according to Yang et al. is 60 to 70 years<sup>11</sup>, and the mean age according to Fanburg-Smith et al is 55 years<sup>8</sup>.

Although rare but have predilection to male children, and head and neck region. They usually come with clinical presentation of an enlarging, rapidly progressive mass with occasional acute hemorrhage, anemia, or coagulopathy<sup>1,3</sup>.

Soft tissue angiosarcoma in children must be differentiated from Kaposi sarcoma, epithelioid hemangioendothelioma, hemangiopericytoma, and spindle cell hemangioendothelioma whose prognosis is different<sup>4</sup>. Also some reports show a frequent mistake of diagnosing as Hemangioma<sup>9</sup>. In our case also the initial histopathology finding in incisional biopsy was epithelioid hemangioma. Angiosarcoma has been categorized into five broad groups based on a wide range of associated clinical scenarios: lymphedema associated, radiation-induced, post-breast cancer, soft tissue and cutaneous<sup>10</sup>. In our case it was in association with soft tissue. Tumor size is a key prognostic variable for soft tissue sarcomas, and a tumor diameter of 5 cm is generally used as a cutoff for risk-grouping purposes<sup>5</sup>. Mainstay of treatment is complete local excision for localised tumours<sup>1,2,6</sup>. But the role of radiotherapy may be needed for local control<sup>6</sup> whereas role of paclitaxel has been proposed as a single agent monotherapy but its efficacy on angiosarcoma is varying when compared to other vascular soft tissue tumours<sup>5,7</sup>. The overall survival is reported to be between 6 and 66 months, and frequent distant metastasis requiring a new approach to adjuvant regimens<sup>8</sup>. In our case radiotherapy was given and after nine month follow up there was no recurrence.

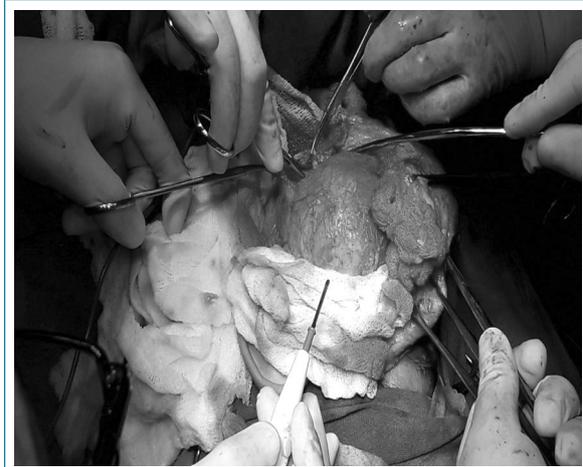


Fig. 4 Peroperative finding.

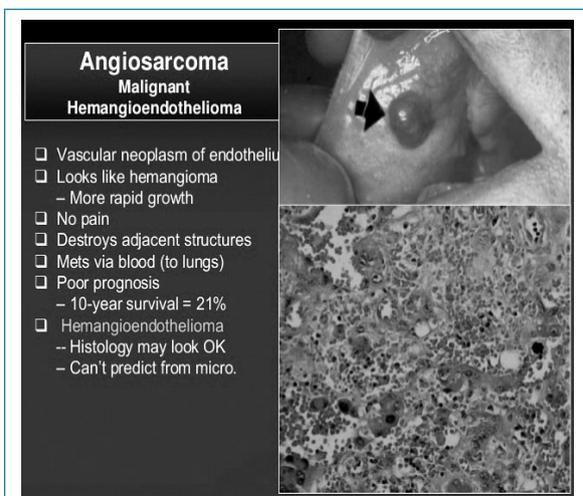


Fig.5 Histopathology finding.

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