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Case Report

Hemangioendothelioma of Lower Lip in A 7 Year Old Boy - A Rare Case Report

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Abstract

The term hemangioendothelioma was introduced by Borrmann in 1899. This tumour was described by WHO as an intermediate malignant neoplasm. Hemangio endothelioma commonly occurs in the superficial or deep soft tissue of the extremities, lungs, liver, bone and lymph nodes, with oral cavity being a rare location. It is usually benign but can show variable grades of malignancy. According to the histological presentation, hemangioendothelioma has been classified as epithelioid, Kaposiform, hobnail (Dabska-Retiform), epithelioid sarcoma like and composite. Here we report a case of hemangioendothelioma in a 7 year old child presented with a complaint of slow growing swelling on right corner of lower lip for 1 year. Its clinical, surgical and histopathological features along with existing literature have been reviewed.

Keywords: Hemangioendothelioma; Lower Lip

Introduction

The term "hemangio endothelioma" was introduced by Borrmann in 1899 who proposed it as an intermediate or low-grade malignant vascular neoplasm. Epithelioid hemangioendothelioma (EHE) was first described by Weiss and Enzinger in 1982 as an angiocentric neoplasm characterized by neoplastic proliferation of cells showing eosinophilic vacuolated cytoplasm and occasional fusiform cells [1] This tumour was described by WHO as an intermediate malignant neoplasm [4]. The lesions of EHE are quite small, superficial, red to brown, angioma-like nodules, usually seen intramucosal or subcutaneously, and are often multifocal. It usually occurs in young adults, with a mean age of 30 years. Lesion often persist but may resolve spontaneously [1]. The tumor is primarily committed to the soft tissues of the extremities; however, cases have been reported in the lung, liver, bone, skin, and head and neck regions, including the oral cavity [3]. The EHE is rare in the head and neck region and even more rare in the oral cavity. Here, we report a case of EHE in a 7year old boy on the lower lip and describing its surgical, histopathological and immunohistochemical findings.

Case Report

A 7 year old boy presented with a complaint of slow growing swelling on right corner of lower lip for last 1 year. Extraoral examination revealed a well-defined growth on the right corner of lower lip of size 2 x 2 cm with a smooth surface. On palpation the lesion was firm, non-tender, well circumscribed with no induration. A provisional diagnosis of fibrosed mucocele or benign connective tissue neoplasm was made. After routine blood examination and screening excisional biopsy was done.

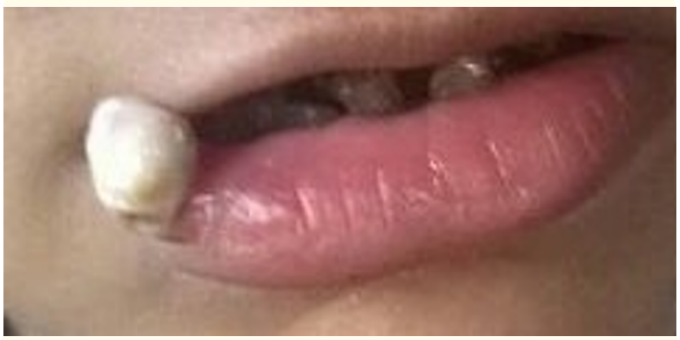

Figure 1: Extraoral photograph.

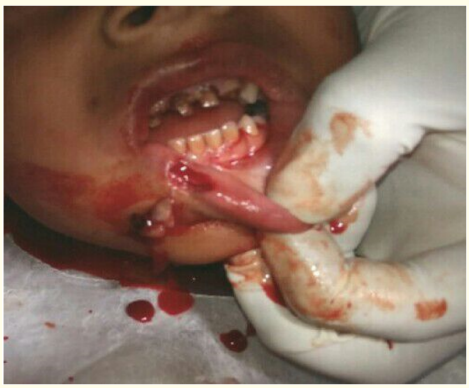

Figure 2: Intraoperative photograph.

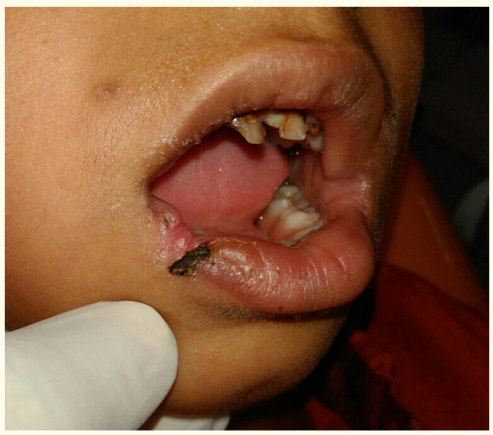

Figure 3: Post-operative photograph after 1 week.

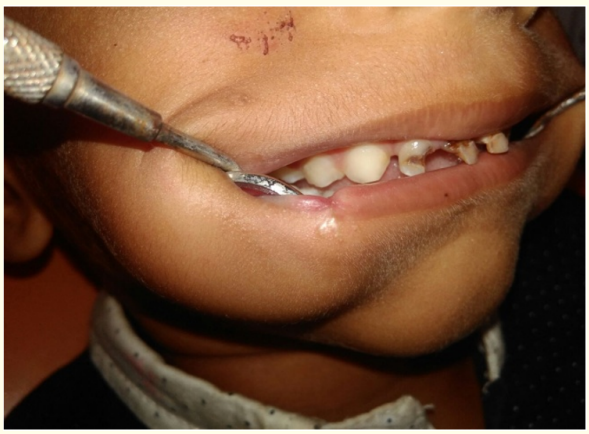

Figure 4: Follow up photograph after 1 month.

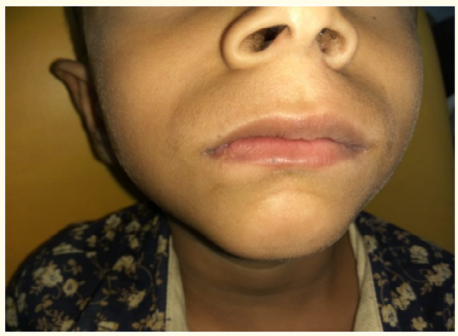

Figure 5: Post-operative photograph after 1 year.

Histopathology

Histopathological examination revealed a parakeratinized stratified squamous epithelium which was ulcerated at areas. Underlying connective tissue was moderately collagenous showing proliferation of vascular channels, epithelioid cells and extravasated red blood cells. Proliferating epithelioid cells appeared plump, with vesicular nucleus, inconspicuous nucleolus, and moderate eosinophilic cytoplasm. occasional mitosis was seen. Diffuse collection of chronic inflammatory cells was also noted. Immunohistochemical staining was performed for CD34. Epitheliod cells were positive for CD34.Based on the histopathological and immunohistochemical features, a final diagnosis of Epitheloid Hemangioendothelioma involving the right side of the lower lip was made. Regular follow-up was done for 1 year and no recurrence was reported.

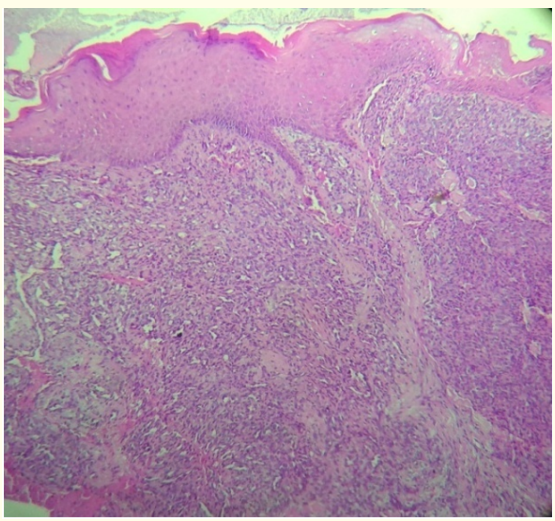

Figure 6: Histopathology (5x).

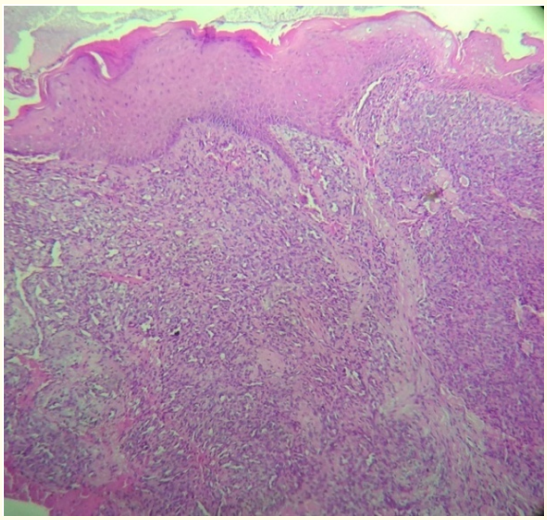

Figure 7: Sheets of endothelial cells in the connective tissue (10x).

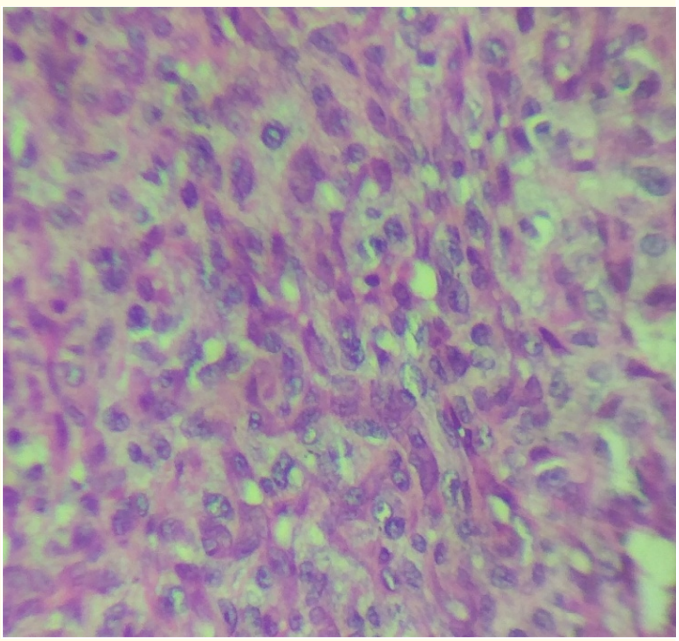

Figure 8: Plump endothelial cells (40x).

Discussion

The term hemangioendothelioma has become a useful designation for vascular tumors that have a biologic behavior intermediate between a hemangioma and a conventional angiosarcoma [5]. Tumors included in this group have the ability to recur locally and have some ability to metastasize, but at a far reduced level compared to angiosarcoma [5].

The epithelioid hemangioendothelioma, an angiocentric vascular tumor, can occur at almost any age but rarely occurs during childhood. It affects the sexes about equally. To date no predisposing factors have been identified [5]. The tumor develops as a solitary, slightly painful mass in either superficial or deep soft tissue, although in rare instances it occurs multifocally in a localized region of the body. At least half of cases are closely associated with or arise from a vessel, usually a vein [5]. In some cases, occlusion of the vessel accounts for more profound symptoms, such as edema or thrombophlebitis [5]. Those tumors that arise from vessels usually have a variegated, white-red color and superficially resemble organizing thrombi, except that they are firmly attached to the surrounding soft tissue [5]. Those that do not arise from vessels are white-gray and offer little hint of their vascular nature on gross inspection. Calcification is occasionally seen in large deeply situated tumors [5].

Pathologically, EHE should be differentiated from other vascular tumours showing epithelioid characteristics, other vascular tumours showing epithelioid characteristics, including epithelioid angiosarcoma, epithelioid hemangioma, spindle cell hemangioma, kaposiform hemangio endothelioma (KHE) and epithelioid angiomatous nodule [8]. Epitheloid angiosarcoma is an infiltrative, destructive vascular tumour, composed of pleomorphic cells, associated with numerous often atypical mitosis, and frequently with necrosis [6]. While a few cases of EHE may show cellular atypia, this lesion does not show the degree of pleomorphism or atypical mitosis seen in epithelioid angiosarcoma. The head and neck region are of particular interest, being the typical location of epithelioid hemangiomas [7].

Epithelioid hemangioma is a circumscribed lesion composed of well-formed, often immature vascular structures lined by plump endothelial cells, in which a large vesicular nucleus often protrudes into the lumina as a _hobnail [8]. This lesion, also coined as angiolymphoid hyperplasia with eosinophilia, for its prominent inflammatory process permits its distinction from EHE [8]. EHE should not be confused with spindle cell hemangioma a benign lesion in older patients with a predilection for the limbs, and characterized by cavernous vascular spaces, with papillary structures, thrombi and phleboliths [8]. Oral carcinoma, which is far more common than EHE, can be relatively easy to differentiate by its sheets of epi-

thelial tumour cells, usually associated with significant pleomorphism, mitotic activity and keratin formation [5].

Reported cases of hemangioendothelioma in the literature

Author (year)	Age	Sex	Site
Wesley., *et al.* (1975)	18	F	Mandibular gingiva+
Ellis and Kratochvil (1986)	13	F	Maxillary gingiva
Ellis and Kratochvil (1986)	4	F	Mandibular gingiva
Moran., *et al.* (1987)	25	F	palate
De Araujo., *et al.* (1987)	4	M	Mandibular gingiva
Marrogi., *et al.* (1991)	45	M	Maxillary gingiva
Marrogi., *et al.* (1991)	36	F	Tongue
Flaitz., *et al.* (1995)	7	F	Mandibular gingiva
Hamakawa., *et al.* (1999)	76	F	Anterior mandible
Orsini., *et al.* (2001)	18	F	Buccal mucosa
Ramer., *et al.* (2001)	32	M	Maxilla
Molina palma., *et al.* (2002)	65	F	Tongue
Machalka., *et al.* (2003)	65	M	Mandible
Anderson., *et al.* (2003)	18	F	Lower lip
Chi., *et al.* (2005)	28	F	Maxillary gingiva
Chi., *et al.* (2005)	23	F	Mandible
Uehara., *et al.* (2006)	72	M	Tongue
Sun., *et al.* (2007)	12	M	Maxillary gingiva
Sun., *et al.* (2007)	53	M	Buccal mucosa
Sun., *et al.* (2007)	17	M	Tongue
Sun., *et al.* (2007)	52	F	Upper lip
Sun., *et al.* (2007)	21	M	Tongue
Sun., *et al.* (2007)	34	M	Tongue
Sun., *et al.* (2007)	11	M	Mandibular gingiva
Sun., *et al.* (2007)	46	M	Tongue
Sun., *et al.* (2007)	6	M	Floor of mouth and tongue
Mohatasham., *et al.* (2008)	9	M	Maxillary gingiva
Robinson., *et al.* (2014)	52	F	Lower lip
Battacharya., *et al.* (2015)	16	M	Maxillary gingiva
Mathew., *et al.* (2017)	30	M	Upper lip
Shijineed., *et al.* (2018)	7	M	Lower lip

Table 1

With regard to therapy, wide local excision was therefore considered the treatment of choice and is probably curative in the majority of cases [9]. The role of adjuvant chemotherapy, radiotherapy, and/or treatment with interferon-a-2 remains unclear [8]. Radiotherapy alone was hardly ever effective because of the slow growth of the tumour cells [8]. Although several chemotherapeutic regimens have been tried, the treatment was not curative [8]. Mortality from the tumour varied greatly, depending on the organ affected and the propensity for multifocality [4]. Reported mortality rates associated with EHEs in more common anatomic locations were as follows: 13% in soft tissue and 35% in the liver [10,11]. Metastasis has been reported in approximately 20% of patients with soft tissue primaries and 25% with liver primaries [8]. On review of the literature, the behaviour of intraoral EHEs appeared less aggressive than those arising from the soft tissues and bones [8].

Conclusion

Epitheliod hemangioendothelioma is very rare in pediatric patients. Only few cases have been reported in the literature. In view of their malignant potential, it appears that wide local excision with close clinical follow-up remains the appropriate management for intraoral EHEs.

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Volume 2 Issue 12 December 2018