CASE REPORT

HAEMANGIOENDOTHELIOMA OF MAXILLARY SINUS: A CASE REPORT

V.P. Narve¹, Aprajita Pandey², Jatin Verma³

HOW TO CITE THIS ARTICLE:

VP Narve, Aprajita Pandey, Jatin Verma. "Haemangioendothelioma of maxillary sinus: a case report". Journal of Evolution of Medical and Dental Sciences 2013; Vol2, Issue 34, August 26; Page: 6561-6565.

ABSTRACT: Haemangioendothelioma is an uncommon neoplasm of vascular tissue in which tumor cells are vascular cells. Its occurrence in head and neck is very rare; only twenty seven cases in English literature have been reported till date. The case described in this report is of a ten year old boy presenting in the ENT Out Patient Department of our tertiary care hospital i.e. G.R. Medical College & J.A Group of Hospitals, Gwalior (M.P) with a maxillary swelling.

KEYWORDS: Haemangioendothelioma, Maxillary sinus, Neoplasm, Vascular cells

INTRODUCTION: The term Haemangioendothelioma has been used over the years for benign and fully malignant vascular tumors composed of endothelial cells, therefore it lacks specificity. Various other pseudonyms have been used in the past, however the term haemangioendothelioma first used by Mallory in 1908 is preferred. Occurrence in head and neck is rare and nose, paranasal sinuses, nasopharynx are relatively uncommon sites for haemangioendothelioma. Rarity of this lesion has prompted us to report this case.

CASE REPORT: A 10 year old Hindu male presented to the outpatient department of our tertiary care hospital i.e. G.R. Medical College & J.A Group of Hospitals, Gwalior (M.P) with swelling over left side of face since 5 months, difficulty in swallowing for 5 months and difficulty in breathing since 3 months.

Fig. I

CASE REPORT

On clinical examination an irregular swelling of 5*4 cm was present on left side of cheek (Fig. I). Swelling was firm, non tender and fixed to underlying structures. Local temperature was not raised. No scar mark or venous prominence was present over surface. Left eye was pushed slightly outwards. No complaint of diplopia was present. In oral cavity a firm, non tender, globular swelling of size 6*5 cm was present. Swelling was firm, fixed, non tender with overlying mucosa smooth, no scar, discharging sinus or pigmentation present on overlying surface. In nose, a reddish mass filling the left nasal cavity was present. It did not bleed on touch and probe could not be passed laterally. Externally dorsum of the nose was deformed and no lymph nodes were palpable in neck. Past history of the patient was not suggestive of any epistaxis or chronic illness. On clinical workup the following picture was present: Total Leucocyte Count - 12,000/mm³, Differential Leucocyte Count - Neutrophils 83, Lymphocytes 15, Monocytes 01, Eosinophils 01, Basophils 00, Haemoglobin 11 gm%, Platelet Count -5 lakhs/mm³, Bleeding Time - 2 min, Clotting Time - 5 min 30sec, Blood Urea - 16mg%, Blood Sugar - 99mg%. On CT Paranasal Sinuses (Fig. II), a heterogeneously enhancing soft tissue lesion with hypodense area suggestive of necrosis with no calcification was present. Expansion of involved sinuses with erosion of overlying anterior, medial and posterolateral wall of left maxillary antrum, hard palate, left sided maxillary alveolar arch and left pterygoid plate was seen. Mass extended to left nasal cavity, left nasopharyngeal and oropharyngeal space, left inferior periorbital fat suggestive of neoplastic etiology. Histopathological correlation was advised.

Fig. II

A punch biopsy via Caldwell-luc approach was initially planned but during the procedure en bloc excision was done and the mass sent for histopathological examination. 1 unit whole blood was transfused postoperatively (Fig. III).

CASE REPORT

Fig. III

Pressure dressing was done, IV antibiotics, anti-inflammatory and analgesics were continued postoperatively and Ryle's tube feeding was given to patient. Rest of the postoperative period was uneventful. HPE report showed vascularised tumor with multiple capillaries.

IMPRESSION: Epitheloid type haemangioendothelioma. Patient was referred to Department of Radiotherapy for further treatment and later lost to follow up.

DISCUSSION: Haemangioendotheliomas belongs to the group of vascular tumors of intermediate malignancy since they are histologically intermediate in appearance between a haemangioma and a conventional angiosarcoma. Another important feature of these tumors is non-homogeneity i.e. each different type exhibits its own unique histological features some times unusual and ambiguous, still displaying all features of a haemangioma, however, with a greater degree of cellularity and mitoses. Not only their histology but also their biological behaviour warrants their separation from haemangioma and angiosarcoma. Fu & Perzin¹ studied non epithelial tumors of nasal cavity, paranasal sinuses and nasopharynx and found the incidence of haemangioendothelioma to be 3.8% out of 81 vascular tumors of these sites. It can occur in the skin, subcutaneous tissue, liver, mammary glands, bones, striated muscles, pleura, uterus, orbit and occasionally tonsils and central nervous system. Maxilla is an uncommon site.

At least three distinctive types of haemangioendotheliomas have been described by Weiss and Enzinger (1985)² i.e. the epitheloid variety, spindle cells variety and malignant endovascular papillary angiosarcoma (Dabska's tumor). Only two cases of haemangioendothelioma of the maxillary sinus have been reported from India in the recent past^{3&4}. A fourth variety, the retiform type has recently been added to the list.⁵

The epitheloid type occurs at variety sites, i.e. skin, bones, lungs, pleura, liver, peritoneum, lymph nodes and rarely in the head neck area.⁶ Most patients are cured by excision but metastasis occurs in one fifth of the cases. Histologically they are composed of distinctive type of endothelial cells having epithelial like or histiocyte like appearance, i.e. abundant eosinophilic cytoplasm with round, vesicular or occasionally indented nucleus. Small vascular lumina are seen. Mitoses, pleomorphism and necrosis are usually scanty or absent. Inflammatory exudate with or without

CASE REPORT

germinal centers may be seen at the periphery. Stroma is scanty or myxoid, occasionally contains osteoclast like multinucleated giant cells.⁷ The endothelial nature of this tumor is confirmed by ultrastructural and immunohistochemical studies.⁸

The spindle cell haemangioendothelioma was first described by Weiss & Enzinger in 1986.² Immunohistochemical features are those of an endothelial tumor.⁹ The third variety, the malignant endovascular papillary angiosarcoma is an extremely rare tumor, occurs in the skin and soft tissue of children.

The retiform type of haemangioendothelioma, a low-grade variant for angiosarcoma, usually occurs in the distal extremities of the young.

In view of the low grade nature of this tumor it has been suggested that complete and wide local excision even without adjuvant radiotherapy or chemotherapy should be the preferred mode of treatment. For histologically malignant forms with regional lymph node metastasis a radical neck dissection should be considered. Radiotherapy should be reserved for multifocal and recurrent disease and the choice of treatment when bleeding is imminent.¹⁰

REFERENCES:

1. Fu Y.S. and Perzin K.H. Non epithelial tumors of the nasal cavity, paranasal sinuses and nasopharynx: a clinicopathological study. 1. General features and vascular tumors. Cancer, 1973,33:125
2. Weiss S.W., Enzinger F.M. Soft tissue tumors- St. Louis, Mosby. Haemangioendothelioma: Vascular tumors of intermediate malignancy. 1989,533-544.
3. Singhal Ashok, Yadav R.C., Kulkarni A., Singhal A.K. Haemangioendothelioma of maxilla, Ind J Otolaryngol 199, 42:73-74.
4. Seth L.S., Bhargava S.K., Bhaya M.H., Ogale S.B. Spindle cell haemangioendothelioma of the maxillary sinus. Ind J Otolaryngol and Head and Neck Surgery, 1993, 2: 211- 213.
5. Calonje E., Fletcher COM, Wilson Jones E., Rosai J.: Retiform haemangioendothelioma. A distinctive form of low grade angiosarcoma delineated in a series of 15 cases. Am. J. Surg. Patho/1994, 18(2): 115-125.
6. Weiss S.W., Ishak K.G., Dail D.H., Sweet D.E., Enzinger F.M. Epitheloid haemangioendotheliomas and related lesions Semin Diagn Patho 1986,3:259-287.
7. Williams S.B., Butler B.C., Gilkey F.W., Kapadia S.B., Burton D.M. Epitheloid haemangioendothelioma, with osteoclastic giant cells, Arch Pathol Lab Med 1993, 117: 315-318.
8. Arnold, G., Klein P.J., Fischer R. Epitheloid haemangioendothelioma,. Report of a case with immunoelectin histochemical and ultrastructural demonstration of its vascular nature. Virchows Arch (A), 1986, 408:4
9. Fukunaga M., Ushigome S., Nikaido T., Ischikawa E., Nakamori K. Spindle cell haemangioendothelioma. An immunohistochemical and flow cytometric study of six cases. PatholInt 1995, 45: 589-595.
10. Taylor G. Etheredge S.N. Haemangioma of mandible and maxilla presenting as surgical emergencies. Am J Surg1984, 108: 574.

CASE REPORT

AUTHORS:

1. V.P. Narve
2. Aprajita Pandey
3. Jatin Verma

PARTICULARS OF CONTRIBUTORS:

1. Associate Professor & HOD, Department of ENT, G.R. Medical College & J.A Group of Hospitals, Gwalior, M.P.
2. Resident, Post Graduate Student, Department of ENT, G.R. Medical College & J.A Group of Hospitals, Gwalior, M.P.
3. Resident, Post Graduate Student, Department of ENT, G.R. Medical College & J.A Group of Hospitals, Gwalior, M.P.

NAME ADDRESS EMAIL ID OF THE CORRESPONDING AUTHOR:

Dr. V.P. Narve,
Associate Professor & HOD,
Department of ENT,
G.R. Medical College & J.A Group of Hospitals,
Gwalior, M.P.
Email- drvpnarve@gmail.com

Date of Submission: 22/07/2013.
Date of Peer Review: 23/07/2013.
Date of Acceptance: 19/08/2013.
Date of Publishing: 23/08/2013