Massive Cavernous Hemangioma in Facial region since 57 years: A rare case report

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***Abstract*- Introduction: Cavernous hemangiomas are composed of large, dilated vascular channels. Compared to capillary hemangiomas, cavernous hemangiomas are more infiltrative, frequently involve deep structures, and do not spontaneously regress.**
Case Report: A 57 year old patient presented in Surgery OPD with a complaint of swelling on right cheek since birth, with the swelling was now increasing in size and tender. On MRI, a large polypoidal exophytic mass of size 7.6x9.5x15cm (APxMLxSI) was seen arising from skin and subcutaneous plane of right cheek. MRI gave presumptive diagnosis of Plexiform Neurofibroma, and histopathological correlation was advised.
Gross examination: examination: Specimen received consist of three greyish brown to greyish black, soft to firm tissue pieces partially covered with skin.
Microscopy: Histological features were consistent with Cavernous Hemangioma.
Discussion: This is probably the first reported case of such massive facial cavernous hemangioma in adult male. This case is particularly interesting because of its presence in an adult male while most of the haemangiomas involutes by 9 years of age.

INTRODUCTION:

Cavernous hemangiomas are composed of large, dilated vascular channels. Compared to capillary hemangiomas, cavernous hemangiomas are more infiltrative, frequently involve deep structures, and do not spontaneously regress. More often, the tumors are of little clinical significance, but they can be cosmetically troublesome and are vulnerable to ulceration and bleeding.
Cavernous hemangiomas constitute one component of Von Hippel Lindau Syndrome, in which vascular lesions are commonly found in the cerebellum, brain stem, retina, pancreas and liver.
In some cases, cerebral cavernous hemangiomas are familial, caused by mutations in one of the three genes called CCM1, CCM2 or CCM3.

CASE REPORT:

A 57 year old patient presented in Surgery OPD with a complaint of swelling on right cheek since birth, with diminution of vision in right eye. The swelling was now increasing in size and tender, since 2-3 months. There was no history of trauma. On MRI, a large polypoidal exophytic mass of size 7.6x9.5x15cm (APxMLxSI) was seen arising from skin and subcutaneous plane of right cheek and lateral aspect of right nose involving right premaxillary space and extending from right temporal region near orbit laterally to medial 1/3rd of left half of mandible inferiorly and medially extending superiorly upto inferior margin of right orbit involving preseptal area, however no orbital extension was seen.
The lesion showed multiple flow voids at its origin, however no evidence of facial muscle, extraocular muscles or bony involvement was noted.
Complete Blood Count, Liver function test, Renal Function test were normal.

MRI gave presumptive diagnosis of Plexiform Neurofibroma, and histopathological correlation was advised.

METHODOLOGY

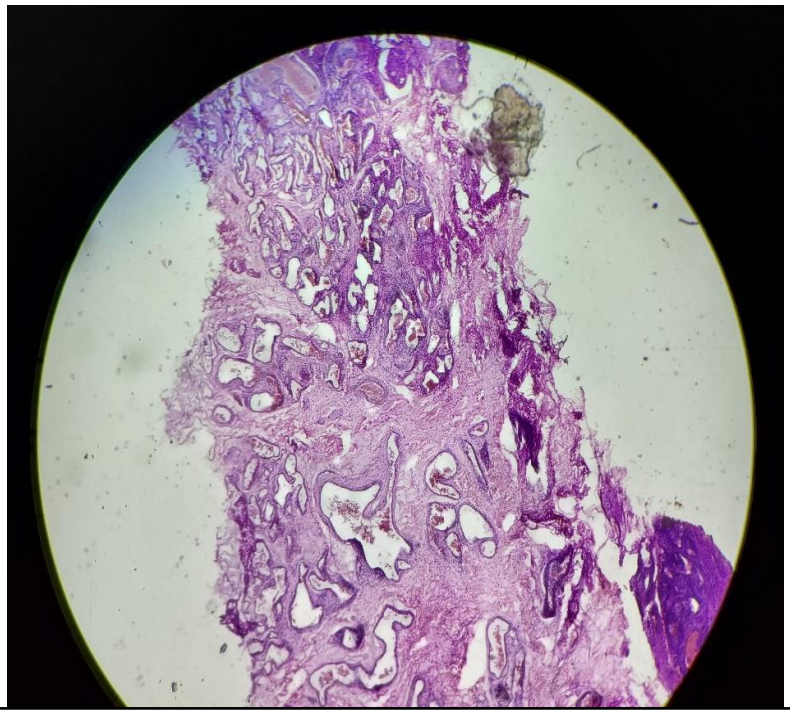
Gross examination: Specimen received consist of three greyish brown to greyish black, soft to firm tissue pieces partially covered with skin. Skin was non ulcerated, normopigmented and non hairy, with multiple variable sized nodular projections seen. Largest piece measured 14x6x3cm, smaller piece 13x6x2.5 and smallest piece 5x3x2cm. On cutting, all tissue pieces were congested and full of blood.

Gross specimen received

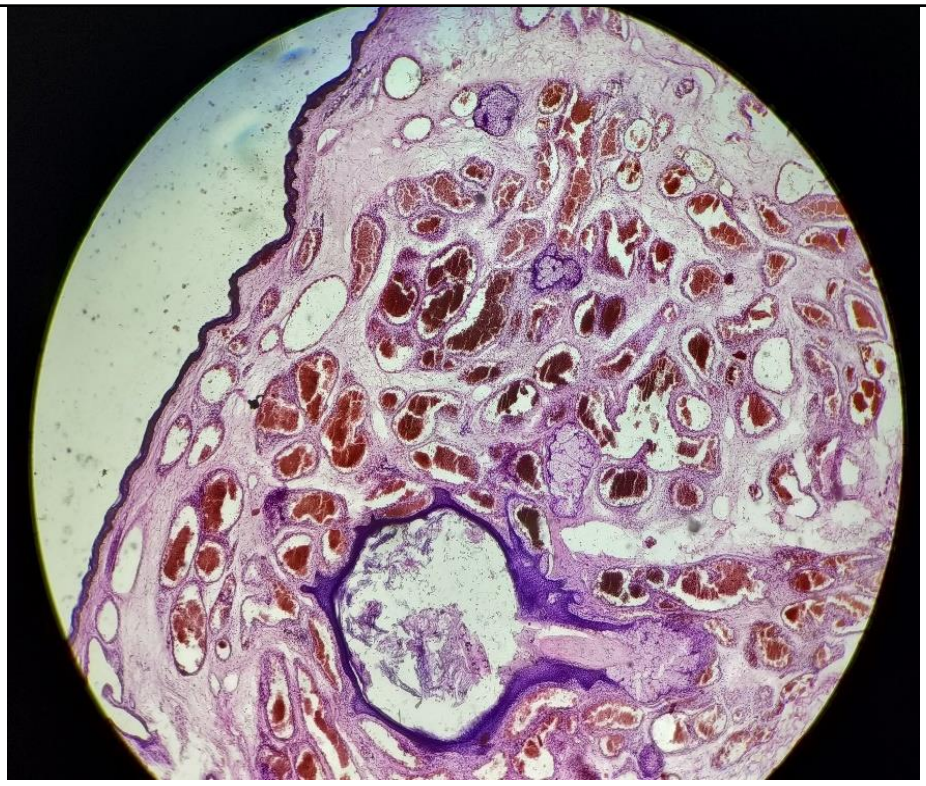
Cut section shows hemorrhagic areas and oozing blood

On cutting, whole tissue was congested

Microscopic Examination: Histological sections showed stratified squamous epithelial lining with subepithelial tissue showing lobules of congested capillary sized vascular channels, lined by single layer of flattened endothelial cells. A large feeding vessel was also seen, with proliferating capillaries seen between the muscle layer.
Histological features were consistent with Cavernous Hemangioma.



20x- Hematoxylin and Eosin stained slide



10x- Hematoxylin and Eosin stained slide

RESULT:
On the basis of Histopathology, the case was diagnosed as Cavernous Hemangioma.

DISCUSSION:
Haemangiomas are a group of vascular tumours, which are rarely present at birth. They have rapid growth during first six months of life and then mostly involutes with full resolution[1]. They are prominent, red and circumscribed lesions with female preponderance (3:1). Commonly, these are isolated lesions (80%), but might be multifocal[2]. The etiopathogenesis of these tumors are unknown, yet there are several theories: congenital, hormonal factors and trauma. There are three types depending on the size of the vessel involved: capillary (small vessels) most common with an estimated incidence of 68%, Cavernous (26%) (Large vessels) followed by compound types (6%). The differential diagnosis includes lipoma, neurofibroma, dermoid cyst, enlarged lymph nodes, soft-tissue sarcoma, myositis ossificans and temporal arteritis[3,4]. Imaging help to make a positive diagnosis and guiding the management of this tumor. magnetic resonance imaging (MRI) is the method of choice in defining the vascular nature of the tumor and providing further information on the exact extent of the tumor. Hemangiomas are generally isointense to muscle on T1-weighted images and hyperintense on T2-weighted images. Several therapeutic modalities are available ranging from simple observation, injection of sclerosing agents, corticosteroid treatment, radiotherapy, embolization (especially in preoperative) to reduce intraoperative bleeding, arriving at complete surgery excision which remains the method of choice in the definitive treatment of this tumor. In some cases of voluminous tumor, injection of sclerosing agents, corticosteroid treatment and radiotherapy, can be indicated as alternatives or adjuvants to surgery[5,6]. The therapeutic indications are made according to the age, the location and size of tumor, growth rate, vascularity, refractory pain, cosmetic malformations, and suspicion of malignancy[7,8].
This is probably the first reported case of such massive facial cavernous hemangioma in adult male. This case is particularly interesting because of its presence in an adult male while most of the haemangiomas involutes by 9 years of age[9].The gigantic size of this facial haemangioma is also not commonly seen as most of them are around 2-4 cm in greatest dimension[10].

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