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Central Haemangioma (Intraosseous) of Jaw

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Abstract:

Vascular neoplasms are common but are often much debated and discussed out entities. These mesenchymal neoplasms are categorized as benign (haemangiomas) or of intermediate behavior (heamangiopericytoma, Hemangioendotheliomas) and malignant as in Angiosarcoma and Kaposi's sarcoma. But even the most benign of entities like haemngiomas can often behave in a nasty way depending on the presentation. Posing great diagnostic dilemma and management delay.We present a rare case of a young boy presenting with an ever increasing firm nodular growth in the outer left mandible. The gingival and the outer skin was intact. Xray suggested of a cystic multiloculated lesion. Biopsy on histopathology revealed an intraosseous haemngioma. A true rarity in itself with extreme paucity of such reported lesions. Again the origins and the cell of origin of haemngiomas have been at major flashpoints and cross firing especially when taken up as differentials with arteriovenous malformations.

***Keyword**: haemangioma, heamngiopericytoma, hemangioendothelioma, angiosarcoma, kapaosi sarcoma, arteriovenous malformations.*

Introduction:

Vascular malformations, vascular hamartomas, vascular tumors are commonly misused interchangeable terms which have come to the foray when one comes across lesions like haemangioma. The term 'Haemangioma" has been misconstrued over time. The nomenclature of haemangioma has been inconsistent over the years and has resulted in complexity while understanding the pathophysiology of tumors. Haemangiomas have ping ponged between tumors and hamartomatous lesions. The origin of hamenagiomas are usually end up with two major hypothesis one which points towards a monoclonal neoplastic proliferation entertaining it as a benign tumor and the other hypothesis stress on the fact that haemangiomas are proliferations of mesodermal cells which differentiate into endothelial cells and subsequently canalize and vascularlize.[1,2] Haemangiomas can be better termed as vasoformative tumors and should be distinctively separated from vascular malformations.[3]

Haemangiomas frequently occur in the first month after birth (almost never at birth) and profusely grow in early infancy only to stop and often regress or partially involute in the next 5 to 7 years after birth. Intraosseous vascular malformations most commonly occur in the fourth decade of life but range from infancy to the eighth decade of life. The peak incidence of central vascular malformations of the jaws is in the second decade of life.[4] Contrastingly, vascular malformation donot follow a parabolic pattern of growth and are rather stable and almost never regress.[3] Haemangiomas commonly occur in the skin and subcutaneous tissues as well as in the organ parenchyma. However haemangiomas of the oral cavity are rare. In the oral cavity the skin, subcutis tissue, muscle and osseous tissue are common sites of involvement. Intraosseous haemnagiomas are a rarity with an incidence of 0.5 to 1.0% of all intraosseous neoplasms.[5] Commonest bones involved are vertebral bodies, calvaria. Amongst the facial bones mandible, the maxilla and the nasal bone invovlment is there.[6]

Central hamenagiomas of the jaw tend to entertain an array of differentials. They are often referred to as 'the great micmickers". They appear on imaging as multilocular cystic lesions commonly giving a kind of honey combing and or a soap bubble apprearance on imaging. Unilocular cystic lesions have also been reported. A point of concern with haemangiomas of intraosseous nature is their existence as a part of a syndrome, e.g. *PHACE(S)* which includes (posterior fossa brain malformations, hemangiomas of the face [large or complex], arterial anomalies, cardiac anomalies, and eye abnormalities and or sternal clefting or supraumbilical raphe). Very rarely the central haemnagiomas may harbor ground for potential malignant transformation.[2]

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Case Report:

This is a case of a 7yr old male child who presents with a left sided madibular swelling since last 3 yrs gradually increasing in size to attain the present size of 7.5x7.0 cms [Fig1]. The swelling is bosselated round with a hard consistency. The swelling is located in the body of the mandible on the left anterior aspect. The lower bony margin of mandible appears to be completely thinned out on clinical examination. Outer skin appears to be perfectly normal with no signs of bruise, ulceration or fistulous tracts. The inner alveolar gingival sulcus as well as the alveolar ridge are perfectly intact with no evidence of ulceration . The teeth in the area around the swelling are intact. On admission the child was subjected to clinical examination for local areas, finds cervical lymph nodes unremarkable (negative for obvious lymphadenopathy). Systemic examination CNS, CVS, RS, GIT within normal limits. The patient has his spleen and liver with normal limits. Xray of the facial bones shows honey combing appearance (multiloculated swelling) suggestive of a cystic

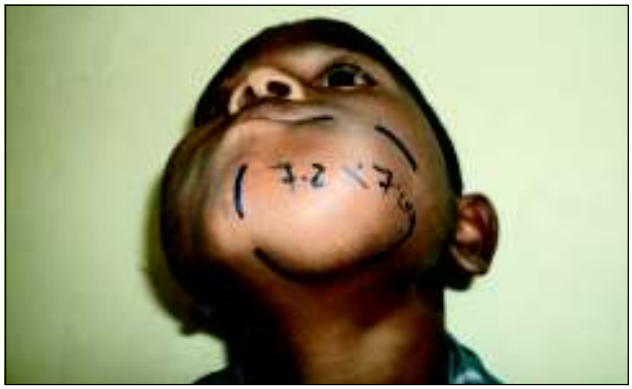

Fig.-1: *showing bosselated nodular firm mass in the left outer mandiblar area*

lesion [Fig 2,3] however, other cystic tumors as well as non tumors pathologies cannot be ruled out (like cystic degeneration in a bone tumor, aneurismal bone cyst, amelobastomas etc) A fine needle aspiration was carried out but turned out to be hemorrhagic and inconclusive. A biopsy was carried out initially reported on histopathology as superficial biopsy and suspicious of fibrous dysplasia, but considering the xray reports the histopathological diagnosis was not conclusive and as a result deeper biopsy (with precautions of a vascular lesion) was carried out which was finally reported as Intraosseous haemangioma (central haemangioma of the Jaw)[Fig 4,5]. A CT angiography was carried out to evaluate in terms of the vascular nature of the tumor and to plan the eventual modality of treatment. Surgical excision was not considered considering the age of the boy and the vascular nature of the tumor. Embolotherapy was carried out to reduce the intraosseous haemangioma. The distortion of the mandible was not a priority in growing child of this age, and the mandible was left intact. On follow up embolotherapy proved ot be extremely beneficial with a drastic reduction in size of the lesion.

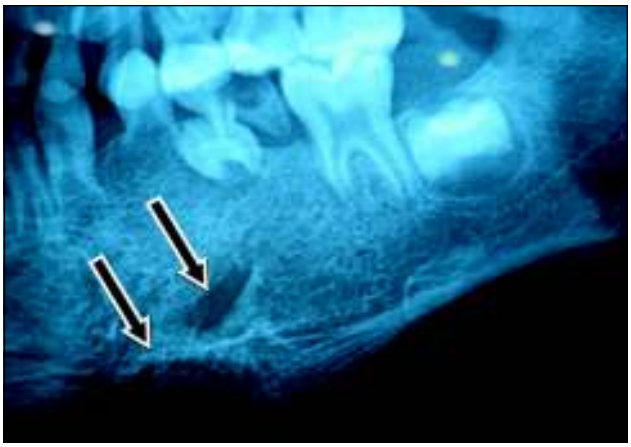

Fig.-2: *X Ray image of the lesion with a honey combing appearance of the intraosseous mandibular lesion.*

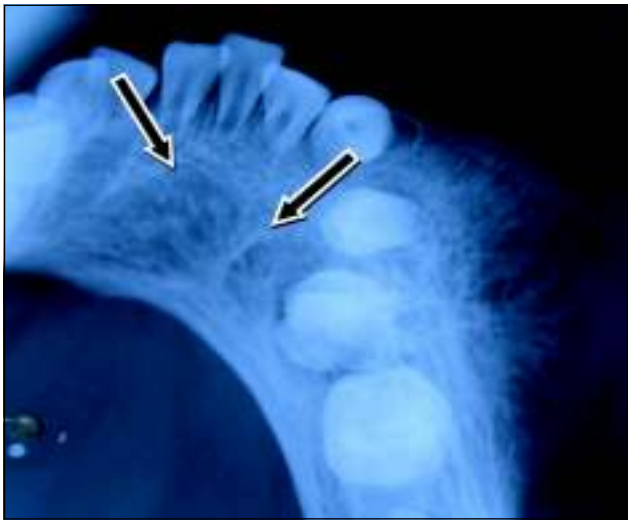

Fig.-3: *X Ray image showing honey combing appearance of the intraosseous mandibular lesion. Suggestive of a cystic loculated vascular origin.*

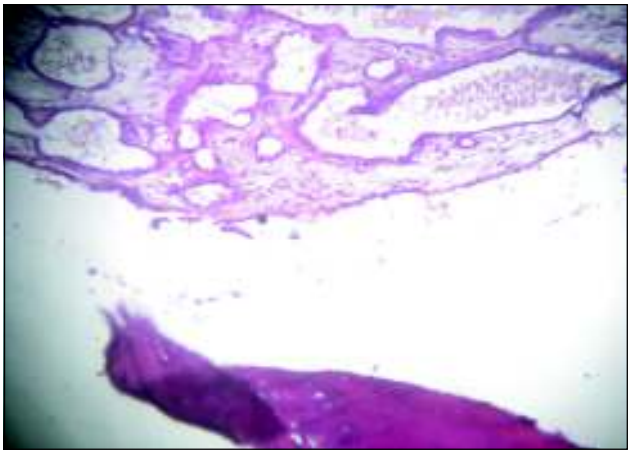

Fig.-4: *H & E stained slide 40x view from the lesion showing proliferation capillary sized vessels, characteristic of a benign vascular neoplasm, Capillary haemangioma, intraosseous*

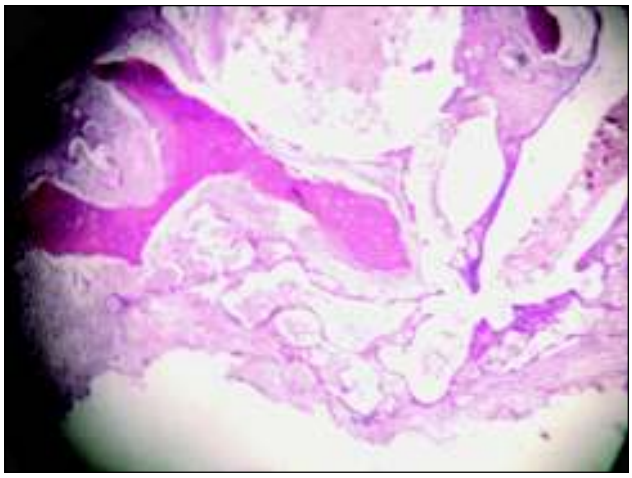

Fig 5: *H & E stained slide 10x view from the lesion showing proliferation capillary sized vessels, along with osseous elements, suggestive of Capillary haemangioma, intraosseous*

Discussion :

This particular case of a 5 yr old boy with a gradually increasing bosselated growth has been interesting in terms of entertaining an array of differentials. At first, clinically thought to be an osseous lesion probably a osteoma or a fibrous dysplasia which was eventually ruled out on imaging studies. The primary histopathological biopsy diagnosis was suspicious towards fibrous dysplasia (though the superficial nature of the biopsy made the diagnosis as suspicious). Another possibility of a Burkitt's lymphoma was not ruled out considering the age and site, however the patient had no 'B' Symptoms and any other complaints implicating a Non hodgkins lymphoma and his blood profile and reticuloendothelial system examination was within normal llimits, negative for any sort of lymphadenopathy, the diagnosis was eventual scraped out. Final a rare diagnosis of central haemangioma (Jaw) was confirmed on histopathological biopsy (second Biopsy). To predict and prognosticate a lesion as central haemangioma it becomes of imperative to ponder and deliberate upon the embryological aspects or the tumorous origins of the same. The remarkable variations shown in terms of site as well as form of presentations of these vascular lesions promote the researchers to put forth two main hypothesis.

Firstly, Researchers point out that the development of haemangioma takes place in three segmental stages in a kind of spectrum. To start with there is an undifferentiated capillary network stage, which invariably is characterized by formation of vascular networking but without differentiation into capillaries of venules or artery., moving on to the retiform stage – this particular stage promotes further enhanced organization of the vascular channels and a better frmae work and cell orientation and lastly the final development stage- this particular stage results in the ultimate cell differentiations from mesenchymal cells to endothelial cells and from supporting fibroblasts to formation of pericytes takes place. With this understanding it can now be stated that the arrest of differentiation at various stages of vascular maturity results in varied outcomes. Say perhaps if we consider the present case, the development of a central heamngioma takes place due to arrest of differentiation from mesodermal cells into undifferentiated capillary networks, and when these arrested meenchymal primordial proliferates it goes deeper into the subcutaneous tissue as well intraosseous and muscular locations. Similarly an arrest in the retiform stage or the final stage of differentiation will result in development of arterivenous malformations and venous - lymphatic malformations respectively. Researches have shown that proliferating haemangiomas have estradiol-17-beta- receptors in their cytoplasm and administration of steroids blocks these receptors.[7]

The second hypothesis states that the cells of origin for vascular development have their precursors located at distant body sites say for example the bone marrow and the placenta. It during embryogenesis and vasculogensis that these precursor cells instead of following the routine vascular pathways to the point of development take a detour and result in vascular malforamtions as well a haemangiomas. This can be stated to be more predominant in during an embolic phenonmenon where in the permissible right to left shunt of fetal circulation may promote such an event. Besides babies of patients undergoing chorionic villi biopsy sampling have a three fold risk of developing haemngiomas which again explains the precursors taking a detour.

Considering the paucity of incidence of central intraosseus haemangiomas, three significant aspects should be entertained when dealing with a potential case of central haemangiomas. Primarily that these tumors can pose themselves as mimicker and so an whole list of differentials should be ruled out based on imaging, like cystic areas - multiloculations as well as uniloculations. Lesions which form close differentials to central haemangiomas include, sarcomas of osseous origin, anneurysmal bone cyst, ameloblastomas, fibrous dysplasia, dentigerous cysts etc. For sake of better understanding and categorizing haemangiomas vs vascular malformations, Mulliken and Glowacki have revised and put forth schematic representations of vasoformative tumors. They have divided the tumors into two broad groups haemangiomas and vascular malformations (arterial, venous, capillary and lymphatic).[3]

Vasoformative Tumor	New Nomenclature	Old Nomenclature
Hemangiomas		
	Capillary hemangioma	Strawberry /Juvenile hemangioma
	Cavernous hemangioma	
	Mixed hemangioma	Parotid hemangioma
Vascular malformations		
	Venous malformation	Cavernous hemangioma/ Hemangiomatosis
	Intramuscular venous malformation	Intramuscular hemangioma
	Capillary malformation	Capillary hemangioma
		Port-wine stain
	Arteriovenous malformation	Arteriovenous hemangioma
		Arterial Cirsoid / Red angioma
		A-V/Serpentine aneurysm

The usual clinical presentation of central heamngioma of jaw, has an history since almost after birth of a jaw swelling which has an indolent behaviuor ever increasing in size, hard in consistency and nontender. However optimal utilization of imaging studies to rule out haemangioma (intraosseous) as a diagnosis, should be considered before any sort of surgical biopsies. The imaging becomes particularly handful aid, so as to plan a proper intervention modality. Cautiously preventing a catastrophic bleeding eventuality especially when managing an intraosseous haemangioma.[8]

Secondly the fact the haemangioma of central intraosseous character is a rare presentation such scenarios should always have an through clinical evalution and elaborate laboratory profiling to rule out multisystem invovlment. Possibilites of syndromic associataion are to be considered.

Hemangiomas are associated with the following syndromes:

1. **Rendu-Osler-Weber syndrome** (autosomal dominant inheritance, multiple telangiectasias, occasional GI tract involvement, occasional CNS involvement)
2. **Sturge-Weber-Dimitri syndrome** (noninherited and nonfamilial, port-wine stain, leptomeningeal angiomas)
3. **Kasabach-Merritt syndrome** (thrombocytopenic purpura associated with hemangioma,

   consumptive coagulopathy, microangiopathic hemolysis, intralesional fibrinolysis)
4. **Maffucci syndrome** (hemangiomas of the mucous membranes, dyschondroplasia)
5. **von Hippel-Lindau syndrome** (genetic transmission variable, hemangiomas of the cerebellum or the retina, cysts of the viscera)
6. **Klippel-Trenaunay-Weber syndrome** (port-wine stain, angiomatosis of the extremities)
7. **PHACE(S)** (posterior fossa brain malformations, hemangiomas of the face [large or complex], arterial anomalies, cardiac anomalies, and eye abnormalities): The association is referred to as PHACE(S) when ventral developmental defects, such as sternal clefting or supraumbilical raphe, are present.

The embryology of these tumors bears its importance considering the management part, as most of the haemngiomas may involute with time and those which do not are either rare exceptions or fall into the vascular malformation category. Management extends from symptomatic relief to medical agents as in steroids or beta blockers even embolotherapy can be effective. Final resorts include cryotherapy and surgical excisions.

Conclusion:

This particular case report commands it due record as it is extremely rare in its presentation as well as its diagnosis. Central intraosseous jaw haemangiomas of capillary type are not common. To add to it an oral surgeon has to keep an entire list of differentials when it comes to unilocular, or multilocular cystic lesions on radioimaging. Another aspect of these tumors is propensity to malignant conversions, (though rarely) have to be of kept in mind.

Conflict of Interest : None

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