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Case Report

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A rare case of mesenchymal hamartoma of cheek masquerading as hemangioma

Vivek Viswambharan[1], M. Lakshmi Narayana[1]*, N. Reddy Chaithanya[1], B. N. Kumarguru[2], Bijjam Sushma[1]

[1]Department of Otorhinolaryngology, [2]Department of Pathology, PESIMSR, Kuppam, Chittoor, Andhra Pradesh, India

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***Correspondence:**
Dr. M. Lakshmi Narayana,
E-mail: lakshmi398@gmail.com

ABSTRACT

Hamartomas are non-neoplastic, tumor-like masses characterized by irregular overgrowth in tissues of its origin, resulting from inborn errors that occur during embryonic development and are present from birth. They may develop from any of the three germinal layers of which mesodermal-derived overgrowth is the most common. Hamartomas may be seen anywhere in the body and most often arises from lung, liver, spleen, kidney, gastrointestinal tract, and rarely in the head and neck region. A 23-year-old lady presented with a painless left cheek swelling for three months. On clinical examination, she had a single, firm and ill-defined swelling of size 2×1cms in left cheek. Ultrasound examination showed an ill-defined hypoechoic lesion measuring 2.1×0.8 cms in the intramuscular compartment of left cheek with few cystic areas. On colour doppler there was minimal peripheral vascularity and flow signal suggesting capillary hemangioma. In view of the tumor's vascularity, Fine needle aspiration cytology wasn't performed and she was treated conservatively on oral propranolol. But since no significant response was noted, excision biopsy was done under general anesthesia. The histopathology features were consistent with mesenchymal hamartoma. Post-operative period was uneventful, and she was in regular follow up with no recurrence.

Keywords: Hamartoma, Head and neck, Cheek

INTRODUCTION

The term hamartoma is derived from the Greek word 'Hamartanien' meaning 'to go wrong' or 'Hamartia' meaning defect, error or sin; along with the term 'oma' (tumor-like growth) adapted from 'onkoma or onkos' meaning mass.[1,2] Albrecht was first to use the term hamartoma in 1904 to describe a mass that resembles a tumor but are non-neoplastic malformations or inborn errors of development from any of the three germinal layers.[3] Hamartomas are an abnormal mixture of tissues native to the part from where they arise. These grow along with, and at the same rate as the parent tissue in contrast to the cancerous tumor. They rarely compress or invade the surrounding structures. They may be seen anywhere in the body and most often arises from lung, liver, spleen, kidney, GIT and rarely in the head and neck region.[4] As per literature, head and neck hamartomas have been reported in middle ear, nasopharynx, larynx, tongue, maxilla, gum margins, neck soft tissues and spaces and eustachian tube, but very few have been reported in the cheek.[5-10] We describe a rare case of mesenchymal hamartoma of the left cheek which was masquerading as hemangioma.

CASE REPORT

A 23-year-old lady presented to our ENT department with a painless swelling on the left cheek for three months. Facial examination showed a single, firm and ill-defined swelling of size 2×1 cms in left cheek extending 4 cms anteriorly from the angle of the lip, posteriorly 4

cms from the auricle, inferiorly 2 cms above the lower border of the mandible and superiorly reaching zygoma (Figure 1). Peroral examination showed a firm, ill-defined swelling palpable through the left buccal mucosa and extending superiorly into the upper gingivobuccal sulcus. Ultrasound examination showed an ill-defined hypoechoic lesion measuring 2.1×0.8 cms in the intramuscular compartment of the left cheek with few cystic areas. On color doppler, minimal peripheral vascularity and flow signal were suggesting capillary hemangioma (Figure 2).

Figure 1: (A) Pre-operative picture showing swelling over left cheek; (B) post-operative (3rd-week review) picture with healed scar.

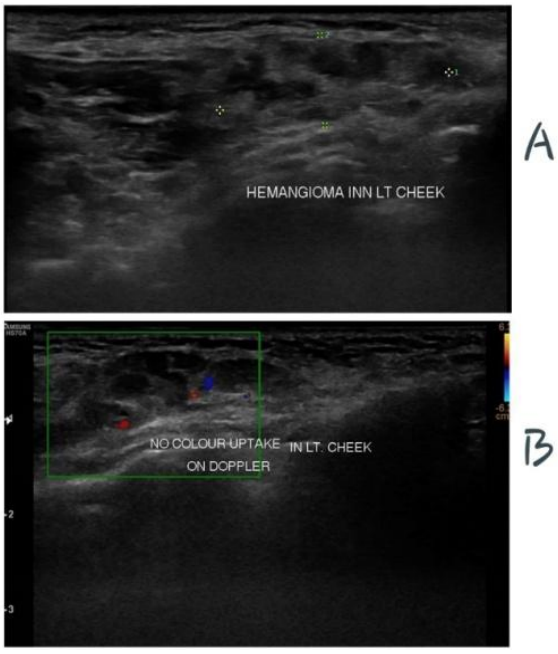

Figure 2: (A) USG showing ill-defined hypoechoic lesion measuring 2.1×0.8 cm; (B) color Doppler showing few cystic areas with flow signal with no color uptake.

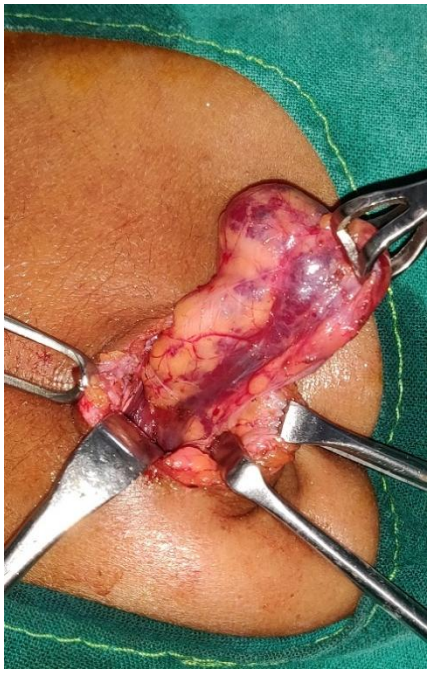

Figure 3: Intraoperative picture showing complete excision of the tumor.

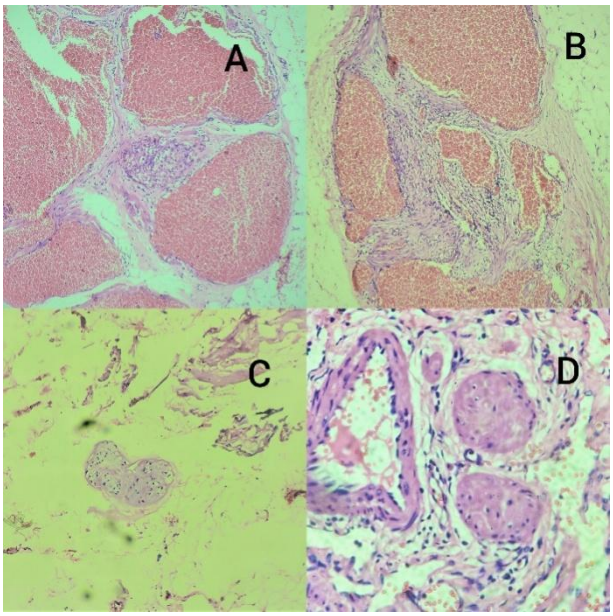

Figure 4: (A) Lobules of capillary sized blood vessels with large cavernous space; (B) dilated and congested blood vessels with cavernous spaces lined by endothelial cells; (C) Islands of cartilaginous tissue composed of chondrocytes and chondroid matrix; (D) congested blood vessels and smooth muscle bundles.

In view of the tumor's vascularity, FNAC wasn't performed and she was started on oral propranolol for three months. Since no significant response was noted, excision biopsy was done under GA (Figure 3). Intraoperatively, we found a lobulated tumor mass with fat and vascular tissue extending below the zygoma and surprisingly not communicating to any major feeding

vessels. The histopathological examination showed lesional tissue composed of adipose tissue displaying numerous large blood vessels arranged in a lobular pattern and lined by flattened endothelial cells and filled with blood. At places, the blood vessels were small and arranged in a lobular pattern. Also seen were bundles of smooth muscle and occasional islands of cartilage, suggesting it as mesenchymal hamartoma (Figure 4). Post-operative period was uneventful, and she was in regular follow up with no recurrence (Figure 1).

DISCUSSION

Gardner discussed the concept of hamartomas and suggested that the term hamartoma should be mainly applied to tumor-like lesions that manifest during the development of the organ or tissue in question, they are mostly non-neoplastic; it should not be used instead of benign tumor.[11] They are often seen in infancy and childhood and thus assumed to be arising as developmental aberrations. It may occur in any organ as it designates a focal overgrowth of mature normal cells and tissues at sites of identical cellular composition.[7] Unlike neoplasms, which are autonomous new growths, the proliferation of hamartomas are self-limiting; they develop until maturity of the tissue is attained and do not exhibit constant unrestrained growth.[12] They do not tend to regress spontaneously either. However, they are prone to recurrences, especially when excised incompletely. These varied characteristics have disputed the neoplastic origin of this uncommon lesion.[13]

The differential diagnosis of cheek swelling in a young female is broad. The benign lesions include vascular malformations, hemangiomas, neurofibromas, teratoma, dermoid, parotid neoplasms, etc. Malignant lesions like parotid cancer and lymphoma too may be considered. Clinically it is difficult to differentiate between teratoma, dermoid, charistomas, and hamartomas.

Hamartomas are classified according to their predominant elements into two types as epithelial (seromucinous, respiratory or salivary) and mesenchymal (angiomatous, chondroid, lipomatous or neurogenic).[14] Histologically, hamartoma must be differentiated from other developmental tumors like teratoma and dermoid.[6] Hamartomas are most commonly derived from mesoderm whereas dermoid is derived from ectoderm & mesoderm and teratoma from all three germinal layers.

Jain et al reported 5 cases of hamartoma of head and neck region involving cheek, neck, maxilla, gum margin and tongue; of which three were of the vascular type, cheek was neurofibromatous type, and maxilla was of sclerosing angiomatous type.[4] In Jain et al study, the cheek case was of an 18-year-old guy who presented with painless, progressive right cheek swelling of size 8×8 cms since birth. In our case, the patient presented similarly with progressive, painless swelling of the left cheek, which she began to notice only for three months. Tanaka et al reported a case of multiple painless nodular swellings over left cheek in a 15-year-old boy with no significant change in size since birth which was then excised and histopathologically shown as striated muscle hamartoma.[15] Al Omran et al reported a case of painless right facial swelling in a five-year-old girl since two months which upon investigating revealed a well-defined ovoid mass within the superficial portion of the parotid gland in CECT which after excision was diagnosed as a vascular hamartoma.[16] For our patient, ultrasound and Doppler were done, which revealed a capillary hemangioma in the intramuscular compartment, and thus we didn't do an FNAC in view of its vascularity. So we started her on medical therapy with oral propranolol for three months. Leaute-labreze et al had reported a complete regression of facial infantile hemangioma in a child who was treated with propranolol.[17] Several authors in their studies have proposed oral propranolol as the treatment modality of choice in case of capillary hemangiomas.

The clinical course of hamartoma is mostly benign, having no tendency to regress spontaneously, and the treatment of choice is complete surgical excision. Nevertheless, there are chances for recurrences, especially when excised incompletely. There are no other documented treatment options for hamartoma and a close clinical follow up is all that is required.

CONCLUSION

Hamartoma should be kept in mind as one of the differential diagnosis for unilateral painless progressive vascular cheek swelling which doesn't respond to propranolol. Hemangioma should be ruled out before arriving at the diagnosis of hamartoma, which is a rarity. Complete surgical excision is the only definite treatment available for hamartoma.

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REFERENCES

1. Ferlito A, Rinaldo A. Developmental lesions of head and neck. In: Barnes L, editor. Surgical pathology of the head and neck. 3rd Ed. Informa Healthcare, USA, Inc. 2009;3.
2. Dehner LP. Congenital anomalies of head and neck. In: Pilch BZ, editor. Head and neck pathology. Lippincott Williams & Wilkins, Philadelphia, PA USA; 2001:103.
3. Albrecht E. Uber Hamartome. Verhandlungen der Deutschen pathologischen Gesellschaft 1904;7:153-7.
4. Jain RK. Hamartoma of the head and neck. Indian J Otolaryngol Head Neck Surg. 1999;51(4):76-8.
5. Baget S, François A, Andrieu-Guitrancourt J, Marie JP, Dehesdin D. Hamartoma of the middle ear: a

case study. Int J Pediatr Otorhinolaryngol. 2003;67(3):287-91.
6. Owens D, Alderson D, Garrido C. Nasopharyngeal hamartoma: importance of routine complete nasal examination. J Laryngol Otol. 2004;118(7):558-60.
7. Windfuhr JP. Laryngeal hamartoma. Acta Otolaryngol 2004;124:301-8.
8. Vashishth A, Mathur NN, Choudhary SR, Khanna G. Giant vascular hamartoma of the tongue. Malays J Med Sci. 2014;21(2):74-7.
9. Bhat KV, Manjunath D, Srinivasan N. Recurrent Lymphovascular Hamartoma of the Neck Spaces. Int J Head Neck Surg. 2006;1(1).
10. Kawamoto A, Katori Y, Honkura Y, Ogura M, Oshima T, Kobayashi T. Osseous hamartoma arising from the eustachian tube. Clin Pract. 2013;3(2):e25.
11. Gardner DG. The concept of hamartomas: Its relevance to the pathogenesis of odontogenic lesions. Oral Surg Oral Med Oral Pathol. 1978;45(6):884-6.
12. Wenig BM, Heffner DK. Respiratory epithelial adenomatoid hamartomas of the sinonasal tract and nasopharynx: a clinicopathologic study of 31 cases. Ann Otol Rhinol Laryngol. 1995;104:639-45.
13. Sekhar LN, Janecka IP (eds). Surgery of cranial base tumors. NY: Raven Press, 1993: 157-223.
14. Carlson ML, Saleh AM, Kaplan KJ, Cofer SA. Mesenchymal hamartomas of the pediatric head and neck. Ear Nose Throat J. 2012;91(7): E6-9.
15. Tanaka A, Hiroi A, Kanehara S, Yamamoto Y, Uede K, Furukawa F. A case of striated muscle hamartoma on the cheek. J Dermatol. 2002;29(11):754-6.
16. Al Omran Y, George SM, Zonoozi S, Al-Omran MK. Vascular hamartoma of the parotid gland. BMJ Case Rep. 2014;2014:bcr2014203615.
17. Léauté-Labrèze C, Dumas de la Roque E, Hubiche T, Boralevi F, Thambo JB, Taïeb A. Propranolol for severe hemangiomas of infancy. N Engl J Med. 2008;358(24):2649-51.

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