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Infantile haemangioendothelioma of the parotid gland: Case report and review of literature

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Abstract

Haemangioendotheliomas (HAE), although rare but are the most common parotid gland tumours in children. We report a 4-month-old girl who presented with a progressively enlarging right sided facial swelling overlying the angle of the mandible. An Ultrasound of the lesion and a computed tomography (CT) scan of the head and neck was carried out which revealed a large lesion within the right parotid gland. CT scan further demonstrated a direct communication with the right external carotid artery and external jugular vein. Considering the clinical course and radiological findings, there was sufficient evidence to avoid any invasive testing. Due to the self-limiting nature of the disease, patient was managed expectantly.

Keywords: Parotid gland, Haemangioendothelioma, Haemangioma.

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Introduction

Haemangioendotheliomas (HAE), although rare but are the most common parotid gland tumours in children.[1-7] They constitute 1-5% of all salivary gland neoplasms.[8] These present in infancy as a painless large rapidly growing mass.[4] It is a benign and self-limiting condition.[4,5] It is therefore of paramount importance that early and accurate diagnosis be made to avoid invasive procedures and alleviate undue anxiety among the caretakers.

We report the case of a infantile haemangioendothelioma of the parotid gland that was managed conservatively at our centre. Although, there have been previous case reports on infantile haemangioendotheliomas,[1-10] this is the first time such a case is coming to light from our part of the world.

Prior permission from the patient's guardians was acquired before the preparation of this manuscript.

Case Report

A 4-month-old girl was brought to the paediatric surgery outpatient clinic on the 22nd of July 2018, with a progressively enlarging right sided facial swelling. Swelling was first noticed at one month age by her parents. It was located at the right side of angle of the mandible and had no overlying erythema or discharge. There was no demonstrable tenderness on palpation. Parents denied any presence of fever, signs of infection or feeding issues and the patient was otherwise healthy with adequate growth and no developmental delay. Patient was born to non-consanguineous parents. She was delivered spontaneously at term without any natal or peri-natal complications. She had no relevant past medical, family or genetic history.

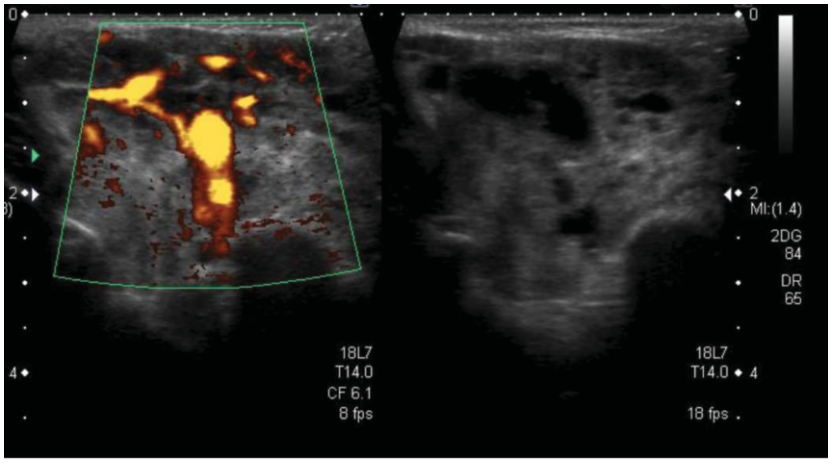

Figure-1: Ultrasound demonstrating internal vascularity in a large parotid swelling.

Investigations ordered showed a haemoglobin (Hb) of 10.5 G/dl, (normal: 9 - 11G a total leukocyte count (TLC) of 14000x109 (N; 5-10,000) and platelets of

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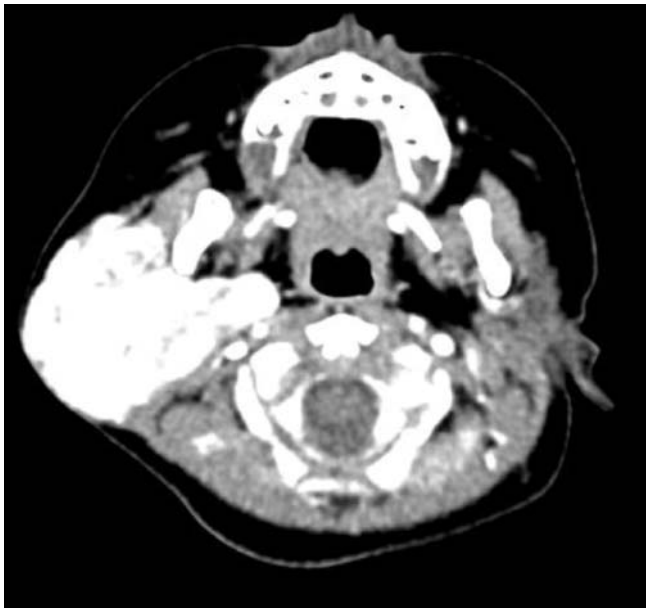

Figure-2: Head CT showing a large right parotid swelling with intense contrast uptake demonstrating vascularity.

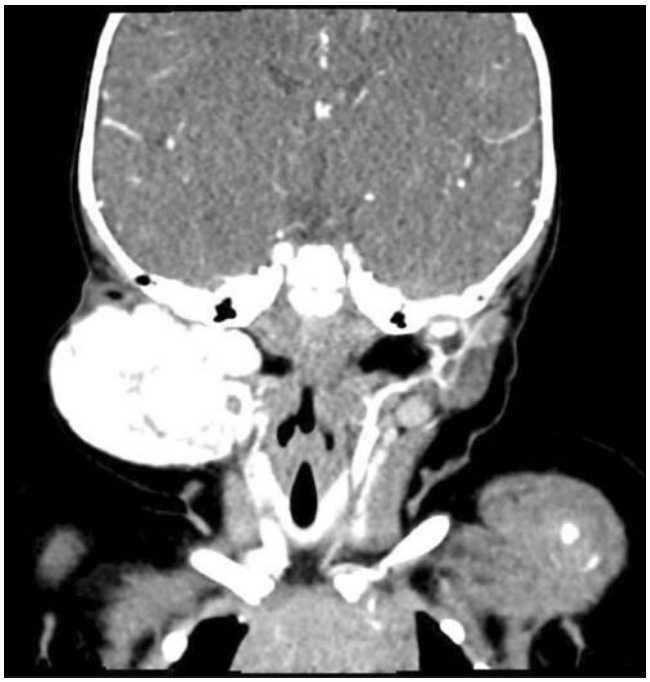

Figure-3: Coronal CT Sequence demonstrating communication of Right Parotid mass with internal carotid artery.

307000x109 (normal: 150-250,000). Ultrasound demonstrated multiple echogenic scattered nodules in the right parotid gland with markedly increased internal vascularity suggestive of a haemangioma. In the light of the above findings a computed tomography (CT) scan of the head and neck was ordered and it demonstrated a large lobulated and vividly enhancing lesion within the right parotid gland measuring 41x43x36mm having a direct communication with the right external carotid artery and external jugular vein. These findings were consistent with a parotid haemangioendothelioma.

Parents were reassured regarding the benign nature of this condition. Patient underwent no further investigations and no therapeutic intervention was deemed necessary. Periodic surveillance for the patient was decided and upon her visit at the outpatient department a month later, no change was noted. Parents were advised to follow up if there were any further development or if any emergent findings were evident.

Discussion

Haemangioendotheliomas (HAE) are neoplastic growths of vascular endothelial cells.[11] Juvenile haemangiomas may occur on any part of the body but most commonly occur in the head and neck region particularly, the parotid gland.[6] These are benign in nature and most cases of HAE appear in the early months of the life.[5] Median age of the presentation is around 4 months.[1,3,4] There is a female predominance with a 3:1 ratio compared to males.[3,4,8] Bluish/purplish discolouration of the overlying skin is a suggestive clinical sign.[1,8] The diagnosis may also be supported by a cutaneous strawberry haemangioma either locally or elsewhere.[5,8]

Infantile parotid haemangiomas have a natural history that includes rapid growth during infancy followed by gradual involution at around 8-18 months.[1,4] They are usually not noticeable in the neonatal period but become prominent in the first few months of life.[1,5]

Characteristic imaging findings aid diagnosis so as to avoid invasive biopsies in the infants. Sonographic findings include a homogenous mass arising in the parotid gland with a lobular structure, fine echogenic internal structure and a lobulated contour.[7,8] The presence of large blood vessels within the tumour is highly suggestive.[4] CT scan shows a soft tissue mass that enhances with contrast material. On MRI these lesions appear isointense when compared to muscle on T1-weighted MRI and they show hyperintense or intermediate signal on T2-weighted MRI. The lesions exhibit homogenous contrast enhancement on MRI.[11] MRI is the best imaging modality.[1,3]

Table: Case reports on infantile haemangioendothelioma of the parotid gland, published in the last 10 years that were accessible as full text, English articles.

Author	Clinical features	Imaging findings
Chaubal et al, 2017[4]	4 month old infant with painless swelling on left angle of mandible	USG: Homogenous, isoechoic lesion with lobulated margins replacing parotid gland. Few echogenic septae seen Colour Doppler showed multiple dilated vascular channels MRI: Homogenous contrast enhancement
John et al, 2016[8]	3 month old female with rapidly enlarging left parotid swelling and cutaneous strawberry haemangioma on the face	USG: Enlarged parotid gland with hypoechoic and lobulated contours. Multiple large anechoic vascular channels noted within. Colour Doppler showed high density of blood vessels within. CT: Non contrast images showed diffuse enlargement of superficial and deep lobes of parotid with lobulated contour. Post contrast images showed intense enhancement of the entire left parotid gland.
Chatura et al, 2015[2]	5 month old male with 2 month history of right facial swelling. Parotid resected due to suspicion of cystic hygroma. Pathological findings suggested of arteriovenous haemangioma associated with CMV infection	USG: Large lobulated tumour
Kotrashetti et al, 2015[1]	Two and a half month old baby boy with 15 days history of left parotid swelling that was gradually increasing in size	USG: Enlarged parotid (3.1 x 2.2 x 2.5 cm) with increased vascularity MRI: well-defined, lobulated homogenously enhancing lesion diffusely involving the superficial and deep layers of the parotid gland
Tandon et al, 2011[7]	3 month old male with right sided swelling on the face. Swelling gradually increasing in size. Reddish patch on right side of face present since birth (cutaneous strawberry haemangioma).	USG: Large lobulated mass with echogenic septations replacing the right parotid gland Colour Doppler showed multiple intratumoral vessels with both arterial and venous components MRI: lobulated mass with flow voids replacing superficial and deep lobes of right parotid gland. The lesion is isotense to muscle on T1 and hyperintense on T2.
Tuna et al, 2009[3]	4 month old infant in 2 month history of right neck swelling	USG: Colour Doppler showed enlarged and heterogenous right parotid gland MRI: Hyperintense right parotid mass containing vascular flow voids on T2.

Red cell scintigraphy can diagnose head and neck haemangiomas with very high accuracy. It shows uniform, well defined uptake similar to the heart and great vessels.[4]

We came across three cases that reported an association between infantile parotid haemangioendothelioma and a cytomegalovirus (CMV) infection.[2,6,10] This suggests that there might be some aetiological role of CMV in this pathology. However, it is equally possible that these findings may have been purely incidental.

Since, many tumours regress on their own, no active management is required. Surgery is not recommended due to the risk of damage to the underlying facial nerve.[4,8] There is reported success with oral propranolol, it has been proven an effective, safe and well tolerated treatment option.[1,9,12] Propranolol facilitates vasoconstriction, decreases expression of vascular growth factors and activates apoptosis of capillary endothelium cells.[1] Minor side effects include agitated sleep, diarrhoea and hypoglycaemia.[12] These are all easily controlled.[12]

Rare but significant systemic complications of parotid HAE include cardiac failure and Kasabach Merrit syndrome.[4,5] Kasabach Merrit syndrome involves a vascular tumour and developing subsequent thrombocytopenia. Treatment of choice in such cases are oral steroids and interferon alpha.[4,5] Our patient had normal platelet levels, as previously mentioned and so was not currently at risk of developing such a condition.

The Table, summarises case reports published in the last 10 years that were accessible to us as full text English articles.

Conclusion

Suggestive clinical course and characteristic findings on radiology provide sufficient evidence for confident diagnosis of infantile haemangioendotheliomas of the parotid gland. There is no need for invasive biopsy procedures. Prognostically the disease shows resolution with no medical or surgical intervention.

Disclaimer: None to declare.

Conflict of Interest: None to declare.

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