ORIGINAL RESEARCH PAPER
Volume - 13 | Issue - 08 | August - 2024 | PRINT ISSN No. 2277 - 8179 | DOI : 10.36106/ijsr
INTERNATIONAL JOURNAL OF SCIENTIFIC RESEARCH

WHEN A CHEEK SWELLING ISN'T JUST A CHEEK SWELLING: INFANTILE HEMANGIOENDOTHELIOMA CASE REPORT

Paediatrics

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ABSTRACT

Infantile hemangioendothelioma (IH) is the most common parotid gland tumor in children. This case report describes a one-month-old infant presenting with a painless, rapidly growing mass on the right cheek. Clinical examination, ultrasound, and CT scan findings were consistent with IH. Differential diagnoses included parotitis and other benign tumors. The patient was treated with propranolol, resulting in a significant reduction in tumor size. Early diagnosis and appropriate management of infantile hemangioendothelioma are essential to minimize complications and optimize outcomes.

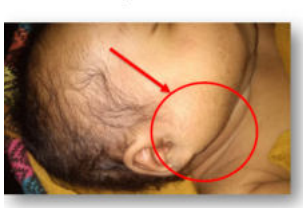
KEYWORDS

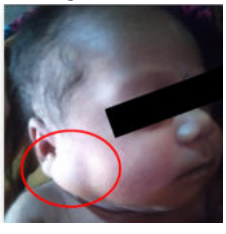
Hemangioma; Parotid gland benign tumor; infantile haemangioendothelioma; Salivary gland tumor

CASE PRESENTATION

A one-month-old male infant presented with a progressively enlarging, painless right cheek mass, first observed by the mother approximately two weeks after an uncomplicated, full-term vaginal delivery. The infant had been exclusively breastfed and was meeting developmental milestones.

On examination, a soft, non-tender, 3 x 3 cm mass was noted overlying the right parotid region. The overlying skin appeared warm with visible vascular dilatation, but without erythema or tenderness. Oral, pharyngeal, and otoscopic examinations were normal. Systemic examination was unremarkable, with normal vital signs.

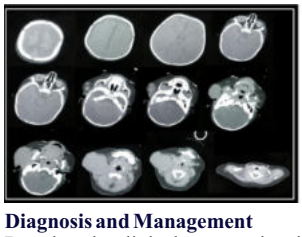


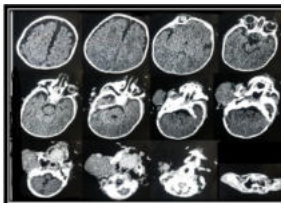


Investigations

Laboratory evaluation revealed mild anemia (hemoglobin 8.3 g/dL), with normal white blood cell and platelet counts. Liver function tests were within normal limits. C-reactive protein was mildly elevated, while amylase and lipase levels were normal.

Ultrasonography of the parotid gland revealed a homogenous enlargement with preserved lobular architecture and prominent vascularity, raising suspicion for a vascular lesion. Contrast-enhanced computed tomography (CECT) of the head and neck confirmed a 41 x 49 x 55 mm multilobulated, intensely enhancing mass within the right parotid gland, extending to the skin laterally and the parapharyngeal space medially.





Diagnosis and Management

Based on the clinical presentation, imaging characteristics, and lack of response to empiric antibiotics, a presumptive diagnosis of infantile hemangioendothelioma was made. The patient was initiated on oral propranolol therapy, resulting in significant regression of the mass, further supporting the diagnosis.

CONCLUSION

This case underscores the importance of considering infantile hemangioendothelioma in the differential diagnosis of a rapidly growing parotid mass in infancy. A comprehensive diagnostic approach, incorporating clinical evaluation, laboratory investigations, and radiologic imaging, is crucial for accurate diagnosis and timely initiation of appropriate management.

Case Discussion

Introduction

Hemangiomas are benign vascular neoplasms that can arise in various tissues, including the brain, kidney, liver, lung, skin, and nasal cavity (5). While rare in the pediatric population, hemangiomas of the salivary gland exhibit a predilection for the parotid gland in young children, particularly females around the age of 4 months (5, 6). Although salivary gland tumors are uncommon in children (<5%), hemangiomas represent the most frequent tumor type in this age group, accounting for over 50% of cases. The vast majority of these tumors originate within the parotid gland (7).

Synonyms (5)

Alternative terms for this entity include:

- Benign Haemangioendothelioma of Salivary Gland
- Immature Capillary Hemangioma of Salivary Gland
- Salivary Gland Hemangioma

Epidemiology

Salivary gland hemangiomas are uncommon, benign vascular tumors that primarily affect the pediatric population. Approximately 65% of these tumors manifest in the head and neck region, with a predilection for the major salivary glands, particularly the parotid gland (5). A study conducted by Weiss et al. revealed that 70% of children with parotid gland hemangiomas were female (6).

Etiology

The precise etiology of salivary gland hemangiomas remains elusive. However, current hypotheses suggest that these tumors arise from vascular malformations occurring during fetal development. Additionally, an association has been observed between these tumors and Kasabach-Merritt syndrome in infants (7).

Clinical Presentation

Hemangioendothelioma (HAE) of the parotid gland, often referred to as infantile hemangioma, is a benign vascular tumor that typically manifests in infancy. It presents as a rapidly expanding mass, frequently undetected at birth but becoming increasingly evident within the first few weeks of life (8). Small lesions are often

asymptomatic, while larger tumors may cause pain and functional impairment due to compression of adjacent structures (8).

On physical examination, HAE characteristically appears as a painless, rapidly growing, well-circumscribed mass. The overlying skin may exhibit a bluish or purplish hue, often described as "strawberry" hemangioma. In some cases, the lesion may extend to involve the cutaneous tissues (8, 9).

Potential Complications

Several complications can arise from parotid gland HAE, including: *(8)*

- Facial disfigurement due to cosmetic concerns
- Functional impairment resulting from compression of surrounding tissues and structures by large lesions
- Ulceration and bleeding at the lesion site
- Facial nerve palsy as a potential complication of surgical intervention
- Post-surgical wound infection

Differential Diagnosis

The differential diagnosis of parotid gland HAE should consider a range of vascular and non-vascular lesions, including:

- Epithelioid hemangioendothelioma (10)
- Low-grade angiosarcoma (10)
- Lymphangioma (10)
- Various subtypes of hemangiomas (spindle cell, capillary, cavernous, hobnail, epithelioid) (10)
- Congenital infantile fibrosarcoma, which may display similar Doppler flow characteristics on ultrasound but typically presents with a heterogeneous internal architecture (11).
- Solitary infantile myofibromatosis, characterized by a period of rapid growth followed by involution, but distinguishable by its hypovascular nature and poorly defined margins (12).
- Sialoblastoma, demonstrating heterogeneous signal intensity on T2-weighted MRI images and contrast enhancement, without evidence of flow voids (13).

Diagnosis

While the clinical presentation often strongly suggests infantile hemangioendothelioma (IH), a definitive diagnosis requires a multifaceted approach encompassing imaging and histopathological assessment.

Imaging Modalities

Magnetic resonance imaging (MRI) is considered the gold standard for evaluating parotid gland masses in infants due to its superior soft tissue contrast and ability to delineate detailed anatomy (14). However, ultrasonography (US) can serve as an initial, less invasive, and cost-effective screening tool. High-frequency US (10 MHz) can often visualize the characteristic features of parotid gland HAE, including a homogeneous, well-circumscribed, lobulated mass with internal septations and prominent vascular structures (14).

Histopathological Confirmation

- Although imaging can provide valuable information, histopathological examination remains essential for definitive diagnosis and to guide treatment decisions. Tissue can be obtained through fine-needle aspiration (FNA) or open biopsy. While FNA is less invasive, it may not always yield sufficient tissue for accurate diagnosis, necessitating an open biopsy in some cases.
- Typically, the biopsy specimen is subjected to hematoxylin and eosin staining, followed by immunohistochemical or molecular studies as needed (7). The presence of reticulum staining is considered a hallmark feature of hemangioendothelioma and confirms the diagnosis (7).

Additional Diagnostic Considerations

In cases with a classic clinical presentation and typical sonographic findings, close clinical follow-up with serial imaging may be sufficient for management. However, atypical sonographic features or uncertain clinical presentations may warrant further investigation with MRI or labeled red cell scintigraphy (15).

In conclusion, accurate and timely diagnosis of parotid gland HAE relies on a comprehensive approach integrating clinical findings, imaging modalities (with emphasis on MRI and US), and histopathological analysis. This multi-modal approach ensures optimal management and improves patient outcomes.

Treatment and Prognosis

- Therapeutic options for infantile hemangiomas (IH) of the parotid gland encompass a spectrum from minimally invasive interventions like cryotherapy and sclerotherapy to more definitive surgical excision, often aided by endoscopy (7). The latter remains the most reliable method for complete tumor removal and prevention of recurrence.
- Pharmacotherapy with oral propranolol has gained recognition as a promising treatment modality for IH. Propranolol exerts its effects through vasoconstriction, inhibition of vascular growth factors, and promotion of apoptosis in capillary endothelial cells (16). Fuschman et al. (17) reported rapid involution of IH at various sites within 2-14 days of initiating propranolol therapy. However, it is important to note that a subset of patients (2-20%) may exhibit resistance to propranolol or have contraindications to its use.
- While most infantile hemangiomas complete their proliferative phase before 9 months of age, some studies have reported recurrence of parotid gland IH following propranolol discontinuation (18). Therefore, a prolonged course of propranolol therapy extending beyond the initial growth phase may be warranted in certain cases.

Prognosis

The prognosis for salivary gland hemangiomas is generally favorable, particularly with appropriate intervention (7). Many cases demonstrate spontaneous regression, obviating the need for surgical intervention. Even in cases requiring treatment, full recovery is often achieved with minimal long-term complications. Conservative management with pharmacotherapy or minimally invasive procedures can often effectively control tumor progression and facilitate regression (7).

CONCLUSION

- Infantile hemangiomas represent the most common parotid gland tumor in the pediatric population. These lesions typically present as painless, rapidly growing masses in early infancy. Clinical assessment, supplemented by imaging studies (particularly MRI), typically establishes the diagnosis (19). Propranolol therapy has emerged as a valuable treatment option, demonstrating efficacy in reducing tumor size and promoting resolution (7). Surgical excision with preservation of the facial nerve remains the definitive treatment for complete tumor removal and prevention of recurrence.

Financial Support And Sponsorship - nil
Conflicts Of Interest - There are no conflicts of interest

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