Case Report

Lipomatosis of the parotid gland in children

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ABSTRACT

Parotid lipomatosis is extremely rare in children. Only 4 cases have previously been reported in the English language medical literature. Surgical excision is frequently complicated by recurrence. We report, a fifth case, on a 5-month-old girl with rapidly progressive parotid lipomatosis. Emphasis is laid on the importance of preserving the unusually delicate tumor capsule to prevent tissue spillage and recurrence. The creation of an appropriate cleavage between the mass and the expanded skin with sparse subcutaneous fat, safeguarding the tumor capsule on one side and the skin blood supply on the other, represents a rewarding technical challenge.

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Parotid gland enlargement arising from infective parotitis is not unheard of in children. In contrast, parotid neoplasms are rare and count for only 1.3% of all benign salivary gland tumors.[1] While Lipomas constitute 2-3% of all benign parotid tumors in children, lipomatosis is extremely rare.[2] Only 4 cases of pediatric lipomatosis of the parotid gland have, hitherto, been reported in the English language medical literature.[3-6] It is a benign, non tumorous condition, defined as deposition of adipose tissue throughout the gland leading to its diffuse enlargement. A fifth case of rapidly progressive parotid lipomatosis in a 5-month-old girl is described, and the condition reviewed.

Case Report. A 5-month-old baby girl, in general good health, was found to have a small nodule in the right cheek, noticed initially at the age of 20 days. It increased progressively in size. Family, maternal, drug, peri and post-natal histories were unremarkable. Examination showed a lobulated, non-tender, irregular, but ostensibly well defined mass measuring nearly 5x6 cm in diameters. It was firm, mobile and apparently not attached to overlying skin or underlying structure. There was no evidence of facial nerve involvement, or lymph nodes enlargement. The computed tomography (CT) scan revealed a soft tissue mass, measuring approximately 4x5 cm, with homogeneous, low-attenuation appearance arising from the surface of the parotid gland, and extending into the pterygoid fossa. A fine needle aspiration biopsy (FNAB) was inconclusive, and the patient was referred to our service.

On examination, clinical finding was confirmed, and a presumptive diagnosis of hemangioma-lipoma was accepted. The patient was kept under observation. Routine laboratory studies such as complete blood cell count, prothrombin time, partial thromboplastin time, random blood sugar, urinalysis and renal function were within normal limits. Alpha-fetoprotein was 44.38 ng/ml (normal: 0-90 ng/ml). The Carcinoembryonic antigen was 2 ng/ml (normal: 0-3 ng/ml). The mass continued to

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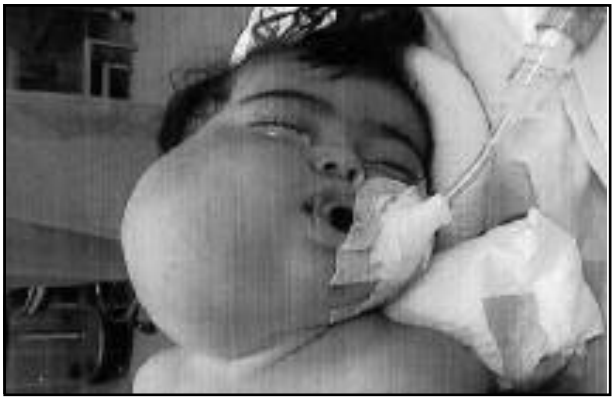

Figure 1 - Infant with a huge mass occupying right parotid area and right side of the neck after endotracheal intubation.

Figure 3 - Intraoperative view showing main trunk of the facial nerve indicated by a right angle clamp, just proximal to its divisions, at final stage of dissection. Note the delicate and transparent capsule engulfing the mass.

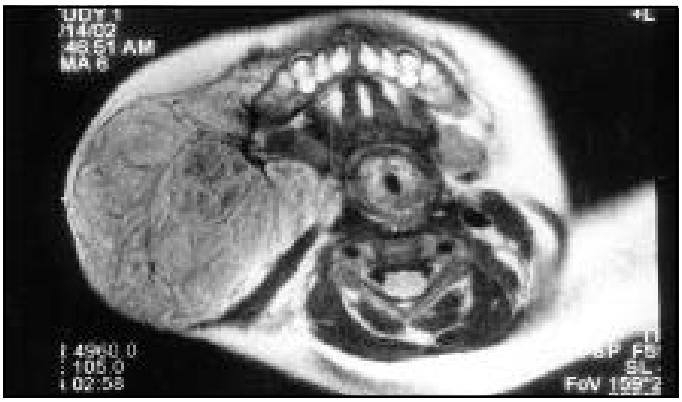

Figure 2 - Magnetic resonance imaging scan depicting the huge mass with an iso to high T2 signal intensity image. Note encroachment of the mass into the parapharyngeal space.

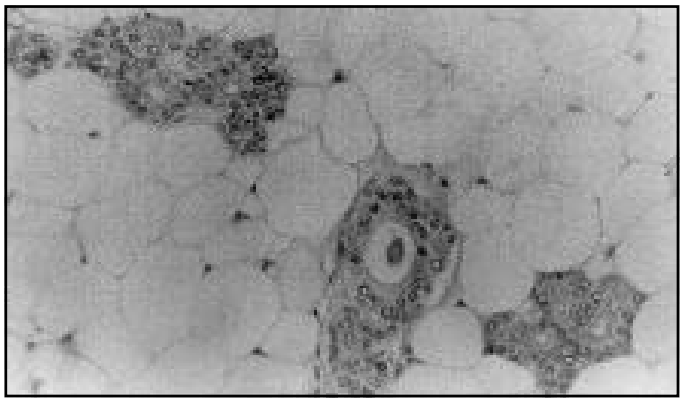

Figure 4 - Extensive replacement of the parotid tissue by mature adipocytes, which separates acini and ducts into small clusters (Hematoxyllin-Eosin x 100).

increase in size to reach the dimensions 10x18 cm, 3 months later **(Figure 1)**. The preoperative magnetic resonance imaging (MRI) showed large, lobulated and well-encapsulated mass lesion in the right cheek and neck, extending into the pterygoid fossa and parapharyngeal space. The signal changes of the lesion were those of fat, confirmed by fat suppression sequence. No local soft tissue or subcutaneous tissue infiltration was noted **(Figure 2)**. A vague diagnosis of a lipomatous mass was made. The differential diagnosis included hemangiolipoma, liposarcoma, pleomorphic adenoma and teratoma.

A classical preauricular incision circumventing the ear lobule and turning forward in a gentle curve to join the upper cervical skin crease was selected. During surgery, a multilobulated, largely lipomatous, well-encapsulated mass, weighing 250 gm, and obviously involving the superficial parotid lobe was identified. This mass was dissected away from the closely neighboring facial nerve, to end up with subtotal parotidectomy **(Figure 3)**. A nerve stimulator was indispensable for this stage of procedure. A special care was necessary to create an acceptable cleavage between the skin flap and the delicate tumor capsule, safeguarding both the structures. At skin closure, the wound margin of the flap, with diminished perfusion, was excised to allow primary wound healing. Histological examination showed well-encapsulated mass depicting diffuse deposition of mature adipose tissue throughout the salivary gland parenchyma without evidence of malignancy **(Figure 4)**.

The infant tolerated the procedure very well. The cosmetic and functional results were excellent. The alpha-fetoprotein level became 8.64 ng/ml 2 months following the operation. There is no evidence of recurrence 7 months after excision and the patient is kept under long term follow up.

Discussion. Lipomatosis of the parotid gland is an extremely rare condition that has first been described in children in 1969 by Johansen and Berdal.[3] It is defined

as diffuse deposition of mature adipocytes throughout the parotid gland, resulting in the separation of acini and ducts into small constellations and islands, under preservation of the basic lobular structures of the gland.[7,8] The condition has grossly been described as a mass infiltrating the parotid gland and the overlying skin.[3] In our case, the mass is identical with the superficial parotid lobe, and separated from the skin by a thin and transparent capsule, clinically appreciable as a distinct margin **(Figure 3)**. One has to differentiate the lipomatosis from an ordinary lipoma, where a discrete and circumscribed lesion is found. It is characterized by the presence of a fibrous capsule, entirely separating the lipomatous tissue from the specific glandular tissue.[4] The absence of lipoblasts enables differentiating lipomatosis from liposarcoma and lipoblastomatosis[7] **(Figure 4)**. We found in the reported cases,[3-6] including ours, 2 patterns of evolution: A rapidly progressive form observed in 3, and a more indolent form in 2 cases. Lipomatosis, in adults, occurs in the neck, trunk, limbs and parotid with similar histopathological appearance. It is found in association with diabetes, liver cirrhosis, chronic alcoholism, malnutrition and hormonal disturbances.[7] Though, there was no evidence of associated ailments in all reported pediatric cases, including this case. As of the rarity of lipomatosis, it is usually not reflected in the preoperative differential diagnosis. It is frequently confused with cystic hygroma, branchial cleft cyst, Warthin's tumor, pleomorphic adenoma and hemangioma.[4-6]

Many imaging techniques, including ultrasonography (US), CT and MRI are available to evaluate salivary gland neoplasms; but none of these can independently give a definite diagnosis. Consequently, a FNAB is considered an essential adjunct to the diagnostic modalities. We think with others that combined US with FNAB are appropriate means to arrive at a definite preoperative diagnosis in the majority of cases of small lesions.[6] However, MRI and even the less desirable CT, due to ionizing radiation, may be required in case of a huge mass to delineate the extent of tissue involvement as demonstrated in this report. The optimal treatment appears to be complete excision, subtotal or total parotidectomy, depending on the extent of the lesion with preservation of the facial nerve. In case of a huge mass, a postauricular incision was suggested to prevent damage to the facial nerve.[4] We found, the classical preauricular incision is adequate for exposure and dissection. Among the 4 reported cases, the excision of the mass was described as having not arrested the disease in a case, perhaps recurrence is intended.[3] Another recurrence is portrayed and accredited to possible microscopic infiltration.[5] We think that the preservation of the unusually delicate tumor capsule may be crucial to avoid tissue spillage and recurrence. However, the high rate of reported recurrence calls for vigilance. The long term follow up is recommended.

References

1. Krolls SO, Trodahl JN, Boyers RC. Salivary gland lesions in children. A survey of 430 cases. ***Cancer*** 1972; 30: 459-469.
2. Walts AE, Perzik SL. Lipomatous lesions of the parotid area. ***Arch Otolaryngol*** 1976; 102: 230-232.
3. Johansen J, Berdal P. Lipomatosis of the parotid gland. ***Acta Otolaryngol*** 1969; 263: 167-169.
4. Adams G, Goycoolea MV, Foster C, Dehner L, Anderson RD. Parotid Lipomatosis in a 2-month-old child. ***Otolaryngol Head Neck Surg*** 1981; 89: 402-405.
5. Holland AJA, Baron-Hay GS, Brennan BA. Parotid lipomatosis. ***J Pediatr Surg*** 1996; 31: 1422-1423.
6. Esposito C, Califano L, D Armiento M, Longo F. Lipomatosis of the parotid gland in a child. ***Br J Plast Surg*** 2000; 53: 699-701.
7. Enzinger FM, Weiss SW. Benign lipomatous tumors, in Soft tissue tumors. St Louis (MO): Mosby; 1983. p. 199-236.
8. Chong GC, Beahrs OH, Chen MLC, Hayles AB. Management of parotid gland tumors in infants and children. ***Mayo Clin Proc*** 1975; 50: 279-283.