

## Parotid Gland Hemangioma

A 5-month-old female infant presented with swelling in the region of both cheeks extending below the jaws, first noticed as a peanut sized swelling 3 months earlier. On examination, there were bright red, bosselated, non tender, non-fluctuant masses in the area of both parotid glands involving most of the cheeks, and both submandibular salivary glands. Overlying skin showed a fine network of capillaries with excoriation on the left side (**Fig. 1, 2**). The infant was born at term, was developmentally normal and growing well. Ultrasonography (USG) revealed enlargement of bilateral parotid, and submandibular salivary glands, lobular structure with fine echogenic internal septations. Color Doppler demonstrated numerous blood vessels within the mass confirming hemangiomas of the glands. Treatment with oral propranolol was started at 2 mg/kg/day in 3 doses; however the patient was lost to follow-up.

Salivary gland tumors are rare in children, most common are hemangiomas and the parotid is most commonly affected. They present as focal lesions or as part of a segmental distribution involving the V3 mandibular segment as in our patient. Segmental

hemangiomas are more aggressive, have a prolonged growth phase, higher likelihood of associated ulceration, airway obstruction, and soft-tissue and cartilage destruction. Diagnosis is confirmed by USG, rarely requires MRI. Lesions to be considered in the differential diagnosis of a parotid hemangioma include cystic lymphatic malformations (cystic hygromas). These do not have a solid component as seen by USG or MRI in PH. Solid lymphatic malformations may extend beyond the parotid and do not contain prominent blood vessels. A rhabdomyosarcoma may arise in the head and neck region, but is rare in infancy, is less vascular and infiltrates into neighbouring structures. Congenital infantile fibrosarcoma of the parotid, though very vascular on Doppler sonography has a non-homogeneous structure. A solitary infantile myofibromatosis in the parotid region may show rapid growth followed by involution, however these are ill defined and not vascular. A sialoblastoma (congenital carcinoma) is a rare tumor of the salivary glands of infancy and can be differentiated from a PH on US or MRI because it is non homogeneous and less vascular.

Most PH involute spontaneously; 10 % require active management. Recommended therapy includes intralesional corticosteroids for small lesions, systemic corticosteroids or interferon alfa-2a or -2b for large ones



**FIG.1** Hemangioma involving left parotid and submandibular glands with superficial excoriation.



**FIG. 2** Hemangioma affecting right parotid gland.

and more recently oral propranolol. Surgical correction is recommended in the involuting or involuted phases (preceded by sclerotherapy).

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## Cerebriform Nevus Sebaceous of Jadassohn

An 11-year-old boy presented with raised pigmented lesion over the scalp since 10 years. Parents gave history of hairless yellowish plaque present over the scalp at birth which gradually increased to present size to take cerebriform appearance. There was no history of trauma. All routine hematological investigations were normal. No systemic and developmental defect was noted. X-ray skull, eye and neurological examination were normal.

On cutaneous examination, single, 17×8 cm brownish, soft, cerebriform and well demarcated nodular plaques was present over the scalp (*Fig. 1*). It had multiple folds. Histopathology showed marked papillomatous epidermal hyperplasia with hyperkeratosis and large numbers of mature sebaceous glands in the dermis along with follicular plugging with malformed hair follicles were also present. The correlation diagnosis of cerebriform type of nevus sebaceous was made.

Nevus sebaceous of Jadassohn (NS) is an epidermal nevus, predominantly congenital sebaceous hamartoma with an estimated incidence of 0.3% in the neonates. PTCH gene deletion is proposed mechanism for development of nevus sebaceous. Cerebriform type is a very rare morphologic variant of NS.

It is usually located over head and neck region as solitary lesion and often present at birth as single hairless yellowish plaque with a smooth velvety surface. Multiple extensive lesions may develop with linear, blaschkoid pattern. It becomes verrucous and nodular at puberty indicating role of hormones. Common sites are scalp, forehead, centropacial, periauricular, and genital area. It may be associated with other developmental defects which are included as epidermal nevus syndrome. Though it occurs sporadically, autosomal dominant transmission was suggested by many case reports. Trichoblastoma is most common benign tumor which develops secondarily in NS, while malignant tumor is basal cell carcinoma (<5%).

The clinical differential diagnosis is congenital



**FIG.1** Cerebriform nevus sebaceous.

melanocytic nevi, epidermal nevus syndrome, giant seborrheic keratosis and warts, while the histopathological differential diagnosis is sebaceous hyperplasia, adenoma, sebaceous carcinoma and sebaceoma.

Seborrheic keratosis and epidermal nevus may be difficult to differentiate clinically. Sebaceous adenomas is sharply demarcated structure made up of immature lobules while in sebaceoma basaloid cells predominate along with sebaceous cells ducts. In contrast to nevus sebaceous, sebaceous hyperplasia shows mature sebaceous gland lobules and prominent sebaceous ductal structures. Sebaceous carcinoma shows mitotic cells with undifferentiated growth.

Wide excision remains treatment of choice and patient mainly present for cosmetic purpose. It may be done prophylactically during childhood as there is a risk of malignant transformation, but most tumors remain benign. So regular clinical follow up is necessary. Many other treatment modalities like CO<sub>2</sub> laser and photodynamic therapy have been tried.

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