Case Report

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Huge Congenital Sialoblastoma

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Abstract

Congenital Sialoblastoma is a very rare salivary gland epithelial tumour. This tumour usually presents in neonatal period or early childhood and can occur in parotid gland or submandibular gland. Sialoblastoma has a potential for local and systemic recurrence, so long term follow up is mandatory. We present a new born male child with a huge submandibular gland sialoblastoma with clinical features, treatment and computerized tomography (CT) findings.

Keywords: Congenital; Sialoblastoma; Submandibular

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Introduction

Sialoblastoma is an epithelial tumour of salivary gland origin and can present in neonatal period or early childhood. Most of the tumours present after the age of 5 years and there is increasing evidence over the first two decades of life [1, 2]. Congenital and neonatal salivary gland tumours are exceptionally rare [3, 4]. Most common site of involvement is parotid gland and only eleven cases have been reported in submandibular gland. Early treatment for sialoblastomas is recommended [5, 6]. We report a huge sialoblastoma in a new-born male child arising from right submandibular region and describe the clinical features, treatment and CT findings of this tumor.

Case Presentation

A one hour new born male child was admitted in our department as a case of huge right submandibular swelling. He was born as a full term neonate by caesarian section. The mass was nodular, firm and fixed to the deeper structures, covered with thin and erythematous skin. Neck, face, right eye, mouth and right ear was distorted by the mass **(Figure 1)**. Computed tomography demonstrated a lobulated mass in the right cervicofascial region. The tumour was arising from the submandibular gland, causing thinning of the mandible, floor of the orbit. Major vessels and veins were well preserved **(Figure 2)**. There was no mediastinal or intracranial extension. Trucut biopsy was done, which revealed sialoblastoma. Complete excision of the mass was done and the patient is doing well in follow up at four months post-surgery. Histopathological examination of the specimen confirmed the diagnosis **(Figure 3)**.

Discussion

Tumours of the salivary gland are rare in children and account for only 3-5% of all tumours [7]. Sialoblastoma was first reported by Vawter and Tefft in 1966 under the name of embyonoma, later on which was named sialoblastoma by Taylor [8, 9]. Developing in the major salivary glands, sialoblastoma has a prediction for the parotid gland and vast majority of cases present in second decade of life [2, 3]. Most of the sialoblastoma present in the neonatal period or early childhood as a neck mass. There is no typical presentation. It is difficult to distinguish these tumors from other more common masses like haemangioma, lymphangioma, vascular malformation, nerve sheath tumors or teratoma. Clinical, radiological and laboratory studies are not difinetly diagnostic for sialoblastoma. An exact diagnosis is made with pathological examination [10]. Characteristically sialoblastoma are composed of numerous solid islands of primitive basaloid epithelial cells and small ducts separated by fibrous to fibromyxoid stroma mimicking the fetal salivary gland. There is peripheral palisading of nuclei in these islands of cells.

Sialoblastoma has generally of a benign nature and total surgical excision is adequate. There is no consensus regarding their biological behavior. It is reported that the tomour is generally

locally aggressive with recurrence rate of 34%. There is no need for chemotherapy and radiotherapy and local excision is adequate even for local recurrence [1-3, 5]. However, this benign behavior is not thought to spread systemically, because it has been reported that sialoblastoma is aggressive and potentially malignant tumour [6, 11]. So the possibility of systemic metastasis should not be forgotten and thus long term follow up is recommended.

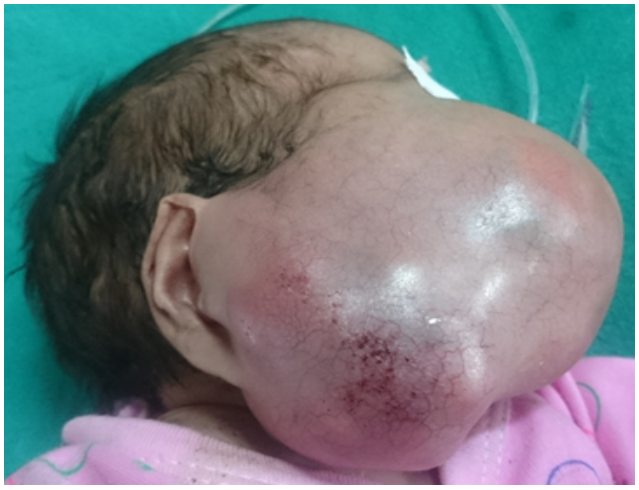

Figure 1 Huge right submandibular swelling causing distortion of the neck, face, right eye, mouth and right ear.

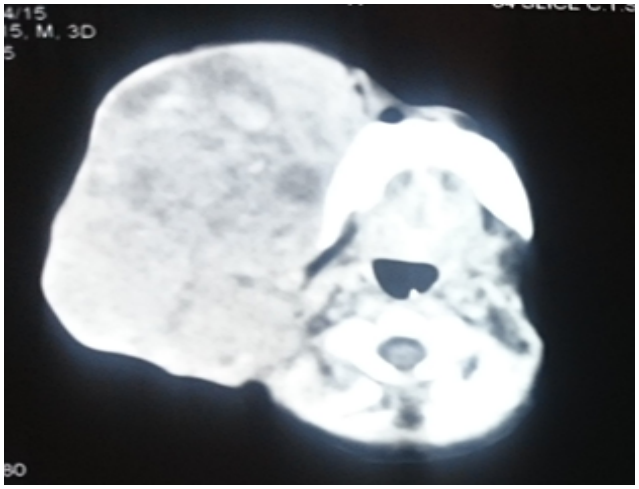

Figure 2 Computed tomography demonstrated a lobulated mass arising from the submandibular gland.

Conclusion

Sialoblastoma is a very rare, aggressive and potentially malignant salivary gland tumour of childhood. Clinical examination, radiological and laboratories studies will only suspect the sialoblastoma and pathological examination is the confirmatory diagnosis. Early surgical excision is prudent and the patient must be followed up for long term to detect any local recurrence or systemic metastasis.

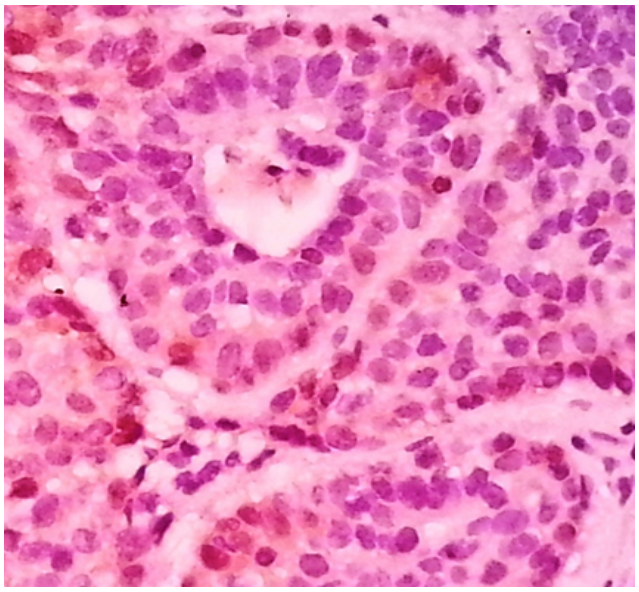

Figure 3 Microscopy of the tumour showing islands of primitive basaloid cells arranged in lobules with ducts in the middle of lobules (H and E).

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