INFANTILE HEMANGIOPERICYTOMA – A CASE REPORT

C. Bharath
Department of Pathology, Medical College (VIMS), Bellary-583104
*Author for Correspondence

ABSTRACT
Background: Hemangiopericytoma is an uncommon and very rare tumor in infancy. This commonly arises on the lower extremities of in retroperitoneum, but occurrence of this tumor in salivary gland is very very rare. Objective: Is that these have a different histological picture and clinical behaviour, when compared to that of adult type. Method: A 3½ months old female infant presented with swelling over right parotid area, which was of 6*3*2 cms in size. A total excision of the parotid gland was done and sent in 10% formalin for histopathological examination. Results: After processing the tissue, multiple sections studied histologically showed numerous capillary channels of varying sizes surrounded by spindle shaped pericytes forming nests and masses, at areas showing mitotic activity and diagnosed as hemangiopericytoma - infantile. Conclusion: Hemangiopericytoma is a very rare tumor in infants and that to the occurrence is very uncommon in parotid region. Rarely these tumors metastasize and may recur locally and behave aggressively with local infiltration.

Key Words: Hemangiopericytoma, Infants, Parotid Gland and Tumor

INTRODUCTION
Hemangiopericytoma is a rare vascular tumor, which was first described by Zimmerman (1923), as pericytes, which is responsible for a vast variety of pathologic behavior patterns relative to different host tissues, rendering an exact assessment of malignant potential of this tumor difficult and confuses the clinician in determining the proper extent of surgical extirpation or use of radiation and chemotherapy. Primary hemangiopericytoma of the major salivary glands are rare (Massarelli et al., 1980). This is an uncommon tumor arising from pericytes of blood vessels. It affects commonly adults with a mean age of 45 years (Enzinger & Weiss, 1995). Occurrence of this tumor in infants is rare. Infantile hemangiopericytoma have a different histological picture and clinical behaviour, compared to adult type. Unlike adult type, mitotic activity and presences of foci of necrosis do not indicate poor prognosis (Enzinger & Weiss, 1995). Occurrence of spontaneous regression has been reported (Chen et al., 1986). These usually follow a benign course and are cured by local excision (Gunawardane et al., 2001). The diagnosis of infantile hemangiopericytoma should be restricted to a lesion that is uniformly pericytomatos and that does not show zonal changes, hyaline or chondroid like nodules or other non-uniform features (Walter & Coulson, 1988). We report the occurrence of infantile hemangiopericytoma in parotid gland, which to our knowledge represents the first one to be reported in this site in South India.

CASE REPORT
A 3½ months old female infant presented with a painless, slow growing swelling in the right parotid gland was admitted to Medical College (VIMS) Hospital, where total parotidectomy was done and sent to our department for histopathological examination.

Pathologic findings
On examination revealed a firm irregular grey brown to grey white mass of 6*3*2 cms in size. On cut section, a grey white tumor was noticed which was around 3*2*1 cms (Fig.1). On microscopic examination, showed sheets of cells which had round to oval nuclei and moderate amount of ill-defined cytoplasm, arranged around variable sized this walled vascular spaces. At areas showed solid sheets of these cells between the salivary ducts. Mitotic figures were rare, necrosis was absent and cellular pleomorphism was minimal (Fig. 2).

DISCUSSION
Hemangiopericytoma arising from or secondarily involving the parotid gland is uncommon to rare neoplasm. Major salivary gland involvement by hemangiopericytoma is almost exclusively an affliction of males. The present reported case being an exception (Carillo et al., 1992). Hemangiopericytoma is a rare tumor which represents only 1.3% of all vasoformative neoplasms of the body (Walke & Bailey, 1971). It may occur in any site, but most commonly in lower extremities (Fischer, 1960) and retroperitoneum. Pagliaro et al., (1988) reported a case of hemangiopericytoma in submandibular gland. The differential diagnosis of infantile hemangiopericytoma includes such as synovial sarcoma, infantile fibrosarcoma
and mesenchymal chondrosarcoma. The infantile fibrosarcoma shows focal biphasic/glandular pattern, hylanisation and calcification, where the infantile hemangiopericytoma lacks these features.

Figure 1: Shows irregular grey brown to grey white capsulated mass, at right below part, cut section showing grey white nodular areas

Figure 2: Histologically showing irregular capillary surrounded by round to oval and spindle like pericytes
(H & E stain. 50x)
Presence of well differentiated areas of cartilage or less frequently bone favours the diagnosis of mesenchymal fibrosarcoma. The range of calibre of vessels seen in hemangiopericytoma is more variable and the spindle cell areas may be confused with aggressive infantile fibromatosis and fibrosarcoma. But the spindle cells in this tumor were not arranged in distinct bundles or fascicles.

CONCLUSION
This rare tumor follows a benign course despite having features which suggests malignancy. A feature which often leads to misdiagnosis of malignancy includes arrangement of cells in solid sheets without characteristic vascular pattern and presence of mitosis and necrosis. Hence extensive sampling will help to identify areas of typical hemangiopericytoma. Rarely these tumors behave aggressively with local infiltration, recurrences and even distant spread (Atkinson et al., 1984).

REFERENCES