PINEOBLASTOMA

CLINICAL DETAILS: -

5-year- female child presented with headache and vomiting for 15 days. No history of fever or seizures. No history of similar complaints int the past. Patient underwent plain and contrast MRI brain in our institution.

IMAGING FINDINGS: -

Plain and contrast MRI brain showed a well circumscribed lobulated solid mass measuring 16x23x22mm (APxTRxCC) with cystic spaces epicentred in the pineal region, indenting on the tectum and extending in to the posterior portion of the 3rd ventricle and aqueduct of sylvius causing mild upstream hydrocephalus. The lesion was heterogeneously iso to hyperintense on T2WI and iso to hypointense on T1WI. Areas of subtle diffusion restriction is seen within the lesion. Scattered areas of peripheral tiny peripheral blooming foci are noted in the lesion. These areas were conferred as calcific foci on post biopsy check CT.

Considering the origin of the lesion with characteristic peripheral burst out calcific foci in a young child imaging diagnosis of Pineaoblastoma was given.

The patient subsequently undergone the transsphenoidal biopsy form the intraventricular component and confirmed to be Pineoblastoma.

DIFFERENTIAL DIAGNOSIS: -

- Papillary tumour of pineal origin- Relatively well circumscribed mass with more of cystic spaces. Calcification is relatively rare. T1 hyperintensity is seen in the cystic spaces due to presence of secretory inclusions containing protein or glycoprotein.
- Pineocytoma- Tumour of adult age group. Well circumscribed mass with homogenous avid postcontrast enhancement. Tumour can show exploded pineal calcification.
- Germinomas- Iso- to hyperintense to gray matter on T1- and T2-weighted images and demonstrate avid, homogeneous enhancement on postcontrast images. The tumours have engulfed pineal calcification.
- Teratoma- Contains central calcification and fat components.
- Ependymoma- Hypoenhancing mass lesion epicentred in the ventricular system.
- Arteriovenous malformation- Illdefined lesion contains a nidus and shows prominent flow voids.
- Choroid plexus papilloma- Relatively well-defined enhancing tumour of young age group with intraventricular origin. Pineal gland is separately disguisable.

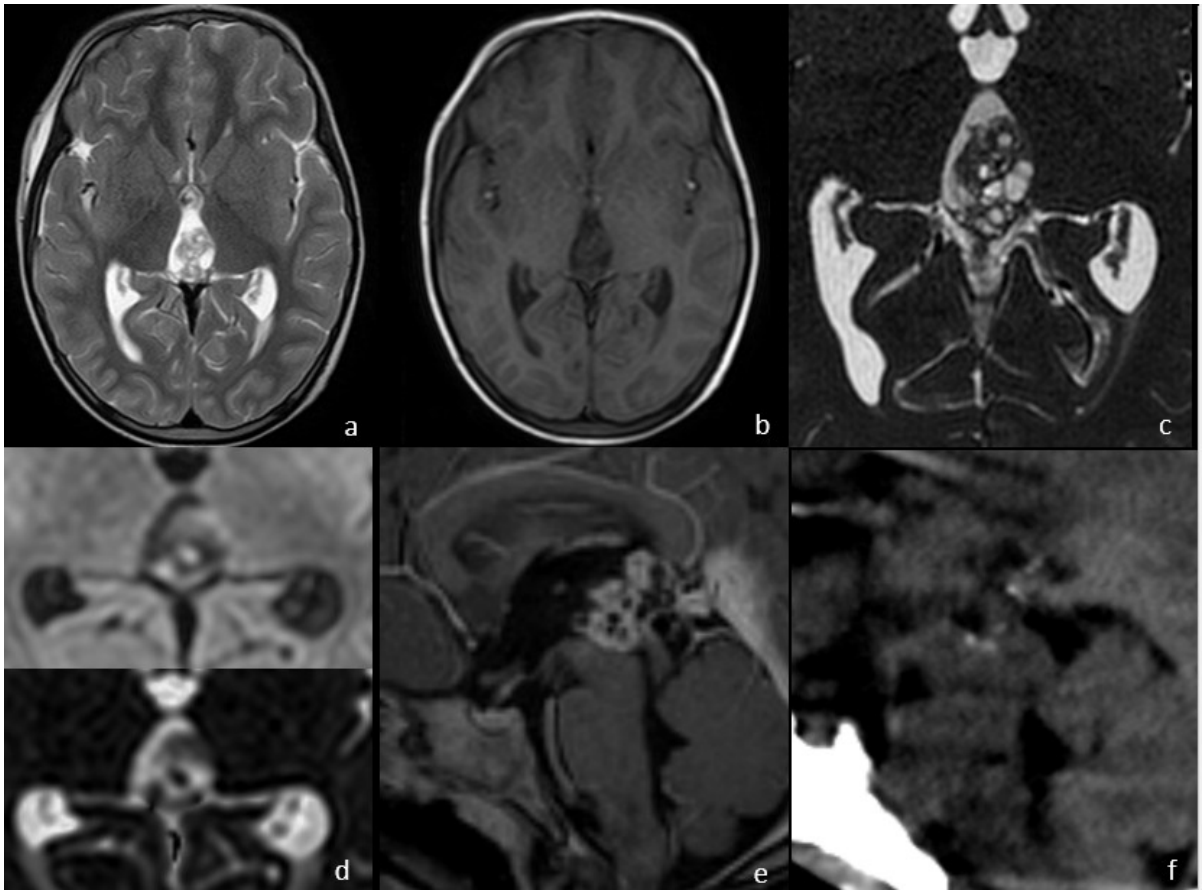

Figure 1: - Axial T2WI(a), T1WI(b) and CISS(c) shows a heterogeneous lobulated lesion in the posterior portion of the 3rd ventricle and aqueduct of sylvius with some cystic spaces within. There is subtle area of diffusion restriction within lesion(d). The lesion has heterogeneous post contrast enhancement with epicentre being the pineal region(d), indenting on tectum and projecting into the ventricular system. Sagittal noon contrast CT section of the pineal region shows foci of diffusion restriction in the periphery of the lesion(e).

DISCUSSION: -

Pineal region neoplasms are more common in children, makes up to 3%–8% of intracranial neoplasms in the pediatric population. Pineal tumours can be broadly categorized int o Primary pineal parenchymal tumours, germ cell tumours, metastasis and lesions arising from adjacent structures. Germ cell tumours are more common and accounts for 40% of primary pineal origin. Pineal parenchymal tumours consist of low grade pineocytoma, intermediated grade-papillary tumour of intermediate differentiation, pineal parenchymal tumour of intermediate differentiation and high grade pineaoblastoma.

Pineoblastoma is a most malignant variety of pineal gland tumour and is classified under WHO grade IV, with a high potential for leptomeningeal spread. They are typically found in young children, with only a slight female predilection. Pineoblastoma when associated with bilateral retinoblastoma, is referred as Trilateral Retinoblastoma. Patients with DICER1 syndrome have an increased risk for developing pineoblastomas.

Almost all cases present with non-communicating hydrocephalus, hence the patients usually present with features of raised intracranial pressure. The tectal plate compression may lead to Parinaud's syndrome.

They are highly cellular tumours resembles primitive neuroectodermal tumor. On non contrast CT they are relatively iso to hyperdense due to high cellularity, with a characteristic exploded calcification in the periphery of the tumour. On MRI heterogenous appearance can be seen with cystic spaces within the lesion. The solid component appearing hypo- to isointense on T1-weighted images and iso- to mildly hyperintense on T2-weighted images. On post contrast images heterogeneous enhancement is observed. Due to its predilection of leptomeningeal spread, whole neuraxial screening is mandatory.

The treatment consists of combination of surgery, chemotherapy and radiotherapy. 5-year survival rate of 60% to 69.5%.

CONCLUSION: -

CT and MRI imaging play a very important role in characterizing the tumours of pineal region which affects its treatment and prognosis.

- Germinomas and Teratomas generally contains engulfed central calcification whereas Pineocytoma, Pineoblastoma and Epidermoid cysts show exploded peripheral calcification.
- Primary pineal origin tumours showing diffusion restrictions are Pineoblastoma, Germinoma and Epidermoid cysts due to its high cellularity. Pineoblastoma generally shows heterogenous contrast enhancement, Germinoma shows homogenous enhancement and Epidermoid cysts lacks internal enhancement.