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Neuroblastoma Presenting as Unilateral Proptosis

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Abstract: ***A 4year old male child presented with unilateral proptosis and fever. CT scan of orbit was performed and an extraconal soft tissue mass was noted in the superior aspect of right orbit with sunburst type of periosteal reaction in the adjacent bone and similar lesions were found in the left orbit and extra axial compartment in median and paramedian region with underlying bone erosion of frontal bone. On performing abdominal examination right flank fullness was felt and following that USG abdomen and CECT abdomen was done. CECT abdomen showed a heterogeneously enhancing mass lesion in the right suprarenal region and our preliminary diagnosis was neuroblastoma. So hereby we are presenting a case of right adrenal neuroblastoma with orbital, intracranial (extra axial) and skeletal metastasis.***

1. Introduction

Neuroblastoma is one of the most common tumours in infancy and childhood to have orbital metastasis.This tumour originates from neural crest cells and most commonly occurs in adrenal gland and also along the sympathetic ganglion chain from the neck to pelvis[1]. Here we aregoing to present a case of right adrenal neuroblastoma with distant metastasis to orbit, intracranial (extra axial), and skull.

Clinical presentation of neuroblastoma mostly will be pain or a palpable mass and abdominal distension, although numerous presentation may be encountered due to local mass effect. In our case the patient presented only with proptosis. Hence proptosis can be the only manifestation of a case of metastatic neuroblastoma.

Although proptosis is not a common complaint in children, its presence usually reflects a serious problem and needs emergent intervention. The risk of malignancies should be considered. Thorough general and regional examination has to be performed with special importance to abdominal examination is mandatory as they may be the only clue for the diagnosis. Early investigation is needed, since the prognosis of the disease is very poor.

2. Case Presentation

4 yr old male child presented with painless progressing swelling of right eye and fever for a duration of 2- 3 weeks.There was no history of visual disturbance and also the patient had discomfortin his right hip. There was no history of past or recent illness or trauma. On local examination there was right side upper lid fullness with proptosis and periorbital ecchymosis (racoon eyes)(fig-1). Rest of the ocular examination was normal bilaterally.

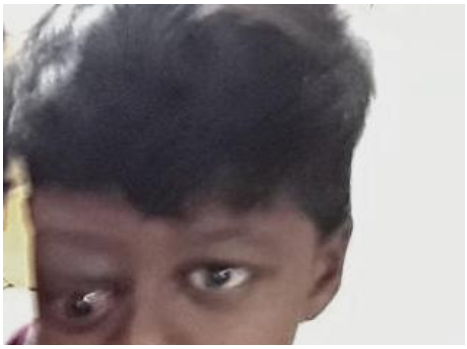


Figure 1

Abdominal examination showed minimal fullness in the right flank. Haemoglobin and haematocrit values were decreased with raised C-reactive protein and erythrocyte sedimentation rate. Ultrasonogram of abdomen was performed and a large heteroechoic mass lesion was seen in the region of right supra renal gland(fig- 2 & 3). Following that CECT orbit and abdomen was performed which demonstrated heterogeneously enhancing mass lesions in the right adrenal gland with lytic lesions in the right iliac wing (fig-4and 5) and superolateral aspect of right orbit with sunburst type of periosteal reactions in the adjacent bone (fig-6 & 7) and extra axial compartment in median and paramedian region with underlying bone erosion of frontal bone (figure-8). Following that bone marrow biopsy was done which confirmed Metastatic small round cell tumour, compatible with neuroblastoma (fig-9).

3. Radiological Imaging

USG ABDOMEN

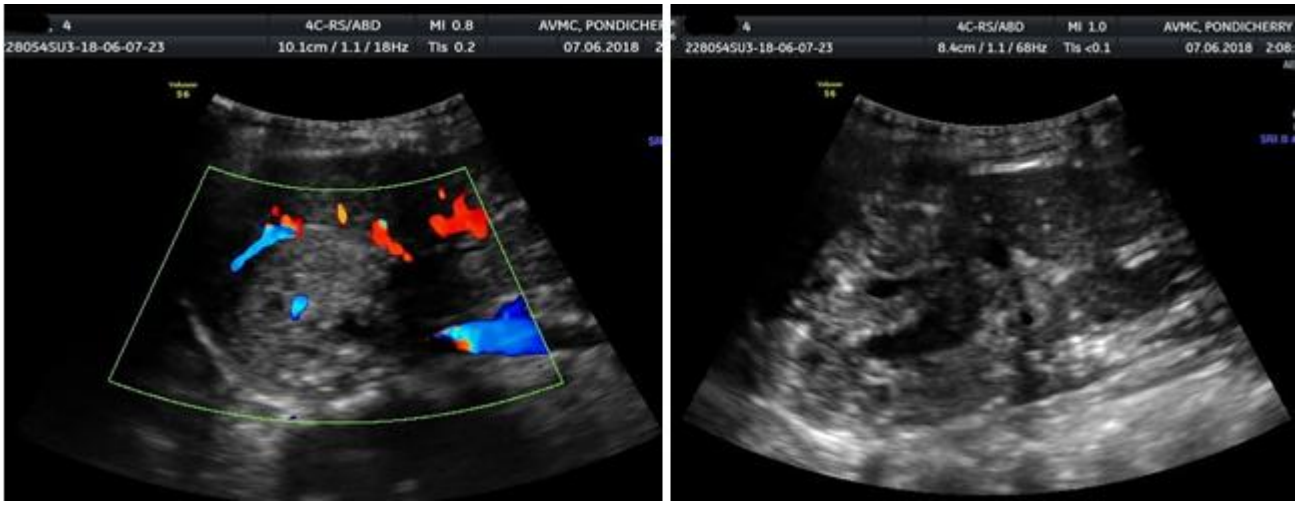
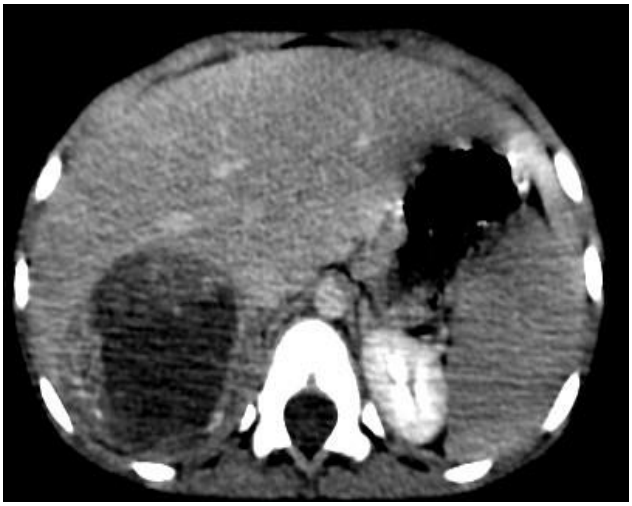


Figure 2 & 3: USG shows a well-defined,heterogenous lesion with significant vascularity seen in the region of right supra renal glandwith few specs of calcifiactions



CECT ABDOMEN- Figure 4: shows a well-defined hetrogenously enhancing lesion seen in the region of right suprarenal gland with few specs of calcification and few non enhancing areas causing mass effect on the right lobe of liver and displacing the right kidney inferiorly

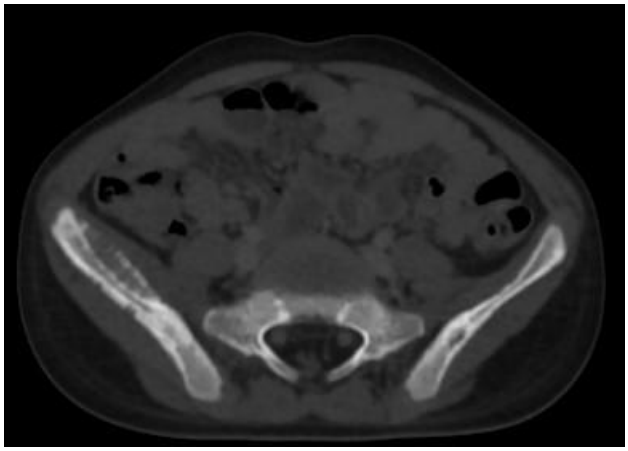


Figure 5: showing lytic lesion in the right iliac wing with sunburst type of periosteal reaction

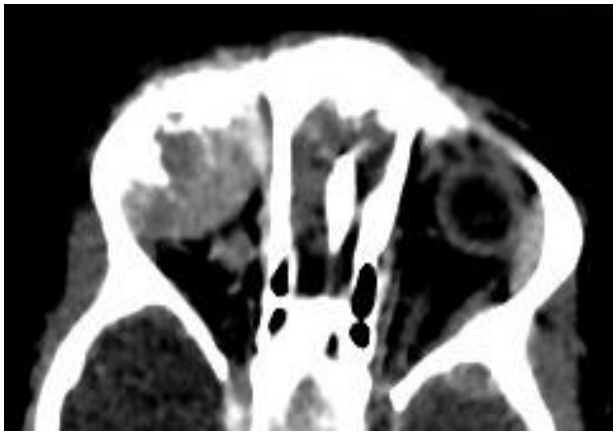


Figure 6: CECT of orbit
Showing a heterogenously enhancing soft tissue density lesion in the superolateral aspect of extra conal compartment of right orbit

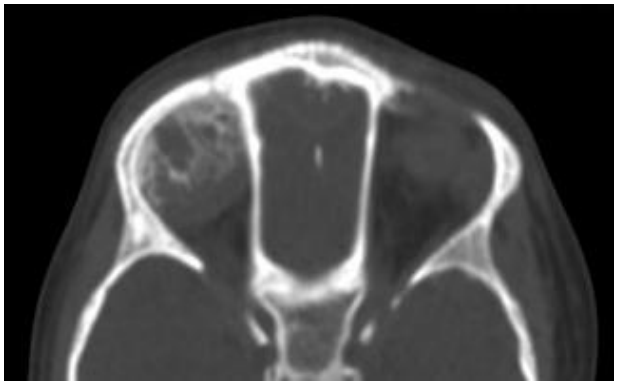


Figure 7: Showing sunburst type of periosteal reaction seen in the roof of the right orbit

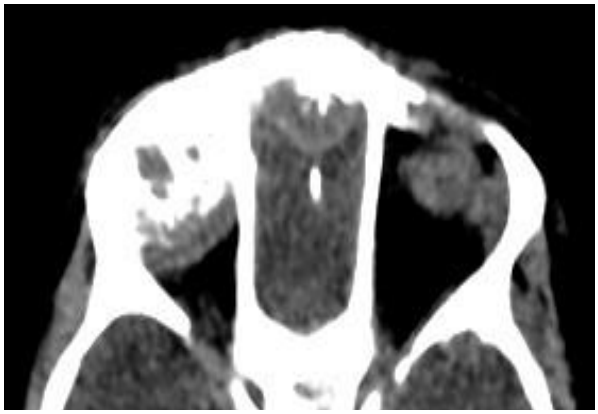


Figure 8: Extra axial lesion with sunburst type of periosteal reaction in the midline of basifrontal lobe.

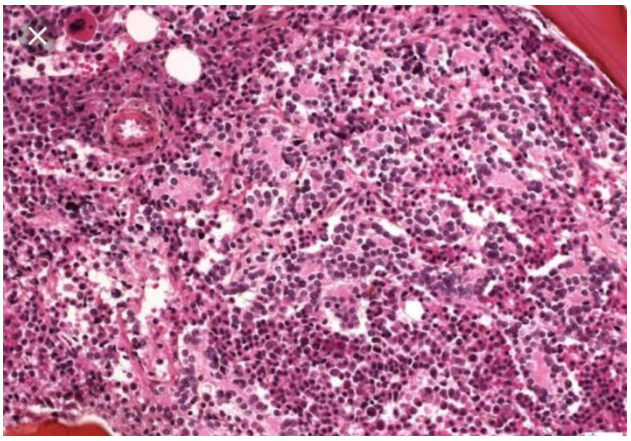


Figure 9: Shows metastatic small round cell tumour, compatible with neuroblastoma

4. Discussion

Neuroblastoma is the most common solid abdominal mass in infancy & the third overall malignancy. Mean age at presentation is 22 months [1]. It arise from neural crest cells and can be found anywhere along the sympathetic chain. Most common site of origin is adrenal gland.Common sites of metastasis is skeletal, liver, lung, pleura, brain, orbit and meninges.Here we have presented a case of neuroblastoma with bilateral orbital, intracranial (extra axial), and skeletal metastasis.

Clinical presentation is with pain or a palpable mass and abdominal distension. Other presentations may be encountered due to local mass effect and metastasis. There are varying accompanying syndromes which include hutchinson syndrome characterised by bone pain or a palpable lump or limping and irritability due to skeletal metastases.[2] Then pepper syndrome which is hepatomegaly due to extensive liver metastasis and blueberry muffin syndrome which represents multiple cutaneous lesions.

CT and MRI are the preferred radiological investigations.The tumour typically appears heterogeneous with calcifications seen in 80-90% of cases [2]. Areas of necrosis are seen as non-enhancing areas. Vascular encasement and mass effect over adjacent organs is noticed. Intraspinal extension of the disease can be better evaluated in MRI.MIBG (metaiodobenzylguanidine labelled to iodine-^{123}) has got significant sensitivity and specificity for detection of neuroblastomassince these tumour cells also secrete catecholamines [2].Bone scan is done with tc^{99}m-methylene diphosphonateis better in detecting and follow-up of skeletal metastases. Our patient was referred to higher centre for further management and on follow up the patients outcome was better.

Treatment and Prognosis

Treatment depends on the patient's stage. Localised tumours are usually considered to be 'low-risk' are surgically excised. In 'high-risk' tumours requires a combination or surgery, chemotherapy with or without bone marrow transplantation is employed, unfortunately with poor overall results. Pre-surgical chemotherapy may also be administered [2].

5. Conclusion

Proptosis can be the only manifestation of a case of metastatic neuroblastoma.Proper local & systemic examination and early investigation is important, as the metastatic disease requires aggressive treatment and the prognosis is very poor.

References

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