Journal of Medical and Biomedical Sciences (2021) 8(1):8 – 11

ORIGINAL ARTICLE

Bilateral Metastatic Orbital Neuroblastoma with Unusual Clinico-radiologic Manifestation

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Neuroblastoma like most primitive neoplasm, primarily affects children under five years. It is uncommon in adolescents and adults. We present a 13-year-old male adolescent with a three-month history of bilateral painful progressive proptosis associated with gradual loss of vision. A contrast-enhanced Computed Tomography scan of the chest and abdomen showed an unusually symmetrical bilateral paravertebral soft tissue masses extending from the cervical to the lumbosacral spines, seamlessly merging with the psoas muscle. A tissue biopsy from the orbital mass reported neuroblastoma. He improved significantly three months following chemotherapy and has been stable during follow up. This case report seeks to demonstrate the many variations in the clinico-radiologic presentation of neuroblastoma and the need for a multidisciplinary approach to patient care, especially in resource-poor setting to ensure good outcome.

Journal of Medical and Biomedical Sciences (2021) 8(1), 8 - 11

Keywords: Bilateral Metastatic Orbital Neuroblastoma, Unusual Clinico-Radiologic Presentation, Ghana

INTRODUCTION

Neuroblastoma (NB) is one of the most prevalent childhood neoplasms. NB represents approximately 8% of malignant neoplasm in children. Its annual prevalence among children in Japan is 150 to 200 (Nakagawara, 2015). NB is relatively frequent in boys compared to girls. Its genetic and epigenetic basis for this prevalence is not clear (Matthay *et al.*, 2016). The prevalence of NB in Japan is comparable to that in the United States of America (USA). NB's phenotype and prevalence depends on race. It is uncommon among blacks but relatively common among whites in North America. Though it is uncommon among persons of African descent, the chances of an aggressive phenotypic malignancy is higher compared to persons of European ancestry (Matthay *et al.*, 2016; Nakagawara *et al.*, 2018).

NB occurrence depends on age. Its incidence peaks among the perinatal age group and gradually reduces during the first decade of life. The disease is uncommon in adolescents and young adults; however, it presents in a more malignant form in this age bracket. NB in children less than18 months of age may regresses spontaneously without any treatment (Matthay *et al.,* 2016; Nakagawara *et al.,* 2018). Paintsil *et al.,* (2015) reported that NB accounted for 4.5% of paediatric cancers in Kumasi, Ghana (Paintsil *et al.*, 2015). The diagnosis should be considered in an acutely ill child presenting with bilateral proptosis with unexplained fever, bone pain and weight loss (Krishnan *et al.,* 2015). This case was a 13 years old male adolescent who presented with bilateral painful proptosis with unusually symmetrical bilateral paravertebral soft tissue masses extending from the cervical to the lumbosacral spines, seamlessly merging with the psoas muscle on contrast enhanced Computed Tomography (CT) scan.

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CASE REPORT

A thirteen years old male adolescent presented to the Tamale Teaching Hospital Eye Clinic with three months history of visual impairment associated with progressive painful bilateral swelling of his eyes. He was chronically sick, cachectic, pale but not jaundiced, afebrile, moderately dehydrated and had no lymphadenopathy. Patient was dyspneic, had stony dull percussion note and decreased breath sounds bilaterally. Heart sounds I and II were present with a galloping rhythm, but no murmurs. He had generalized abdominal tenderness. However, there was no palpable intra-abdominal masses, bone tenderness, weakness or paralysis of the limbs. The vision was 6/18 and 6/60 in the right and left eye respectively, on presentation. There was a total restriction of extraocular movements, proptosis of 30mm right eye and 50mm left eye measured with a ruler from the lateral orbital rim associated with ecchymosis, chymosis and ciliary injection. The cornea was clear and anterior chamber was deep in both eyes. The pupil was round and reacts to light sluggishly in the right eye and there was a relative afferent pupillary defect (RAPD) grade II in the left eye. The lens and vitreous were clear, with no optocilliary shunts or papilloedema in both eyes.

A provisional diagnosis of bilateral orbital inflammatory disease with bilateral pleural effusion to rule out a metastatic malignancy was made. The patient was admitted to the ward and managed initially with broad-spectrum antibiotics and intravenous fluids. Full blood count revealed high white cell counts of $37.21x10^9$/L with neutrophil predominance, hemoglobin level of 10.5g/dl and platelets of $68x10^9$/L. Blood urea, electrolyte and creatinine were within normal limits. A plain chest x-ray revealed bilateral pleural effusion, which was minimal in the left and moderate in the right hemithorax. Computed Tomography (CT) scan with contrast of the head showed a heterogeneous enhancing lesion in both orbits (**Figure 1)**.

A left suprio-lateral orbitotomy was performed and an incisional biopsy taken for histopathology. Left eye temporal tarsorhhaphy was done to protect the conjunctiva and cornea. A right tube thoracostomy was perfumed and it drained approximately 700mls of haemorrhagic fluid. Talc pleurodesis was done after drainage was minimal and lungs expanded. Macroscopically, fragments of tan tissue were received in the laboratory, averaging 0.5cm. Microscopically, sections showed small-undifferentiated cells in sheets with Homer-Wright rosettes, and pseudo-rosettes formation with areas of necrosis (**Figures 2a-c**). A histopathological diagnosis of orbital neuroblastoma was made. The patient was started on oral prednisolone. A CT scan of the chest and abdomen was ordered which showed an unusually symmetrical bilateral paravertebral soft tissue masses extending from the cervical spine to the lumbosacral spine seamlessly merging with the psoas muscle. The chest images showed bilateral pleural effusion more in the right and patchy consolidation in the lower lobes (**Figure 3**).

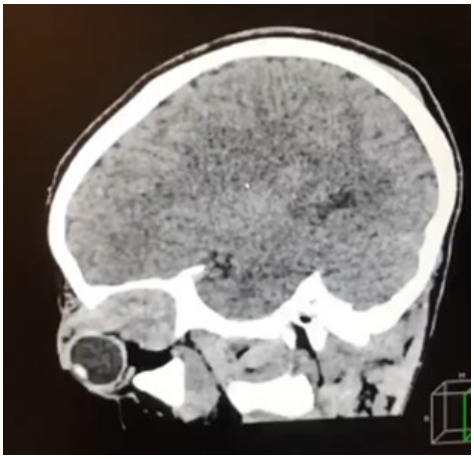

Figure 1: CT Scan with contrast of the head of a 13-year old boy showing a heterogenous retrobulbar mass with uniform enhancement and proptosis of the globe.

A final diagnosis of symmetrical bilateral paravertebral NB with bilateral orbital metastasis was made. Palliative chemotherapy was commenced using a modified protocol for high-risk NB. A cocktail of intravenous vincristine, cyclophosphamide and etoposide were administered. Cisplatin and carboplatin were omitted due to cost. He improved

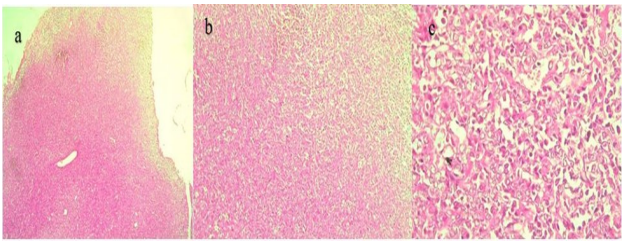

Figure 2: Histological sections from a left orbital mass of a 13-year old adolescent showing dense sheets of small monomorphic primitive cells (X4), a. Small primitive cells with round hyperchromatic nuclei and scanty cytoplasm (X10), b and (X40), c.

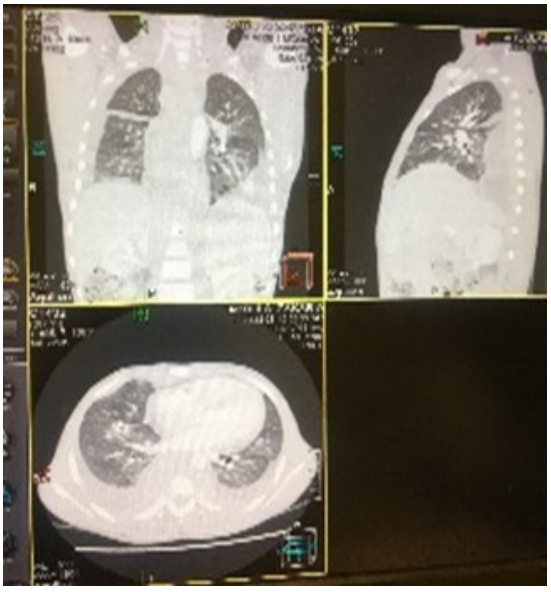

Figure 3: CT Scan with contrast of the chest and abdomen. There is bilateral symmetrical paravertebral masses extending from the cervical to the lumbosacral spine. The chest images showed bilateral pleural effusion with patchy consolidation in the lower lobes.

significantly with reduction of proptosis and resolution of the pleural effusion and was discharged a week later and scheduled for three weekly chemotherapy.

DISCUSSION

Neuroblastoma (NB) is a neoplasm of primitive neuroectodermal cells that originates from neural crest cells. It arises from the sympathetic nervous tissue along any part of the sympathetic chain. (Duarte *et al.*, 2018; Sameer *et al.,* 2017). NB occurrence depends on age. Its incidence peaks among the perinatal age group and gradually reduces during the first decade of life. The disease is uncommon in adolescents and young adults. However, it presents in a more malignant form in this age bracket. In children less than 18 months of age, it may regresses spontaneously with no intervention (Matthay *et al.*, 2016; Nakagawara *et al.,* 2018).

NB has a widely varied clinical course that mainly depends on the biology of the neoplasm. This is directly influenced by the size, location, presence of metastasis, and/or hormonal activity of the neoplasm (Duarte *et al.,* 2018). The presentation of the ocular signs in our patient were consistent with findings reported in other studies where a two year-old female child presented with rapid onset of massive proptosis of the left eye associated with lid edema, ciliary congestion and total restriction of extraocular movements (Vallinayagam *et al.,* 2015). However, our patient was an older male child who presented with late-stage disease. In a study done in Ghana, out of 14 who were diagnosed with NB, eight were male and six female. Only two children were between the ages of 10 to15 years where our patient falls within. (Paintsil *et al.,* 2015).

The radiological features of NB vary widely too, this also depends on size, location, presence of metastasis, and hormonal activity of the neoplasm. On CT scan, NB is commonly detected as a large, lobulated, heterogeneous solid lesion displacing all adjacent structures (Kembhavi *et al.*, 2015). The CT scan of the chest and abdomen of our patient showed an unusually symmetrical bilateral paravertebral soft tissue masses extending from the cervical spine to the lumbosacral spine seamlessly merging with the psoas muscle.

The patient was managed on intravenous vincristine, cyclophosphamide and etoposide. The management of the patient was thus multidisciplinary. However, it was not optimal as cisplatin and carboplatin were omitted due to cost. In General, chemo-reduction is usually done to reduce the size of the neoplasm before surgical intervention

(Kohler, 2013). NB involving the orbit can cause compressive optic neuropathy. Steroids in high doses and/or radiotherapy can prevent this phenomenon. (Rapoport *et al.,* 2020). Oral prednisolone was administered to our patient after the incisional biopsy. The pleural effusion was managed with tube thoracostomy and talc pleurodesis (TP) as recommended for malignant pleural effusion (Shouman *et al.,* 2012; Srour *et al.,* 2013). Effusion resolved and chest tube was removed prior to discharge.

CONCLUSION

Our patient had a more malignant form of NB as an African adolescent and presented with unusual clinico-radiologic features, which compounded the diagnostic dilemma. Nevertheless, he responded very well to the initial treatment. It is therefore, important that one should entertain a high index of suspicion to make an appropriate diagnosis in such cases and institute a multidisciplinary approach in its management to improve survival rates.

COMPETING INTERESTS

The authors declare that they have no competing interests.

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ISSN 2026-6294