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Case Report Open Access

Rare Occurence of Neuroblastoma in Young Adult—A Diagnostic Challenge

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Introduction

Neuroblastoma is a malignant neoplasm derived from the neural crest cells located preferentially at the medullary region of adrenal gland and less frequently at other sites. Case reports of this neoplasm over 18 years of age are rare and this disease shows no preference for gender [1]. Upuntill June 2008 no more than 65 neuroblastomas in adults have been reported in literature [2,3]. The most common location for primary disease in adults are abdomen followed by thorax pelvis and occasionally head and neck [4,5]. Clinical presentation of the disease varies and usually depends on size site of tumour and involvement of adjacent organs or metastatic sites but may occur without any specific symptoms. Biochemical studies have demonstrated 85-90% neuroblastoma patients have elevated catecholamine metabolites in urine but hypertension is rare [6]. Clinical data on survival outcomes of adult patients with neuroblastoma and available information on the influence of the stage biological features and histopathology to their prognosis are scarce due to the rarity of disease.

Case Report

We report a 25 year old male patient presenting to the medical out patient department (OPD) of a tertiary care hospital with symptoms of right hypochondrium pain, vomiting, and palpitations for last 2 weeks. On examination he was anxious and restless, pallor ++, pulse rate 150/min no radio femoral delay, Blood pressure 230/120 mm of Hg. Cardio vascular system examination showed tachycardia, no added sounds and no murmurs. Respiratory examination was normal. Per abdominal examination revealed non tender Hepatomegaly 2 cm below right costal margin otherwise normal. He was admitted in ICCU and then evaluated. His ECG showed sinus tachycardia with heart rate of 160/min. His treatment was initiated with Tab Metaprolol 50mg BID and was kept under observation. On second day his blood pressure was 140/80 mm of Hg HR 100 bpm and he was asymptomatic. We investigated for any secondary cause of labile hypertension in young adult. In the mean process he underwent routine investigations. Hb 7g%, Hematocrit 30%, WBC counts within normal limits; peripheral smear study showed microcytic hypochromic anemia and reticulocyte count was 1%. Blood urea, serum creatinine, serum electrolytes were within normal limits. Urine for VMA, 17 ketosteroids was negative. Hbs Ag and HIV tests were negative. LFT and serum amylase were within normal limits. Chest x ray was normal. Abdominal ultrasound showed fatty liver changes otherwise normal. Renal doppler was normal. He was transfused with 4 units of packed cells. Clinical improvement was noticed and seeing to this he was discharged after 2 days of admission with tab Metaprolol 12.5 mg BID, haematinics and was advised to report to the medical opd after a fortnight.

After 2 months of his discharge this patient was brought to the emergency department with symptoms of colicky abdominal pain, nausea, vomiting, palpita`tions, profuse sweating and restlessness. On examination, he was irritable, pale, significant weight loss of about 25%, with Pulse rate of 140 bpm, blood pressure was 230/120mm of Hg, and ECG showed sinus tachycardia of 150 per minute. Significance was given to severe anemia noticed and progressive weight loss. Now his Hb was 5.2g%, Reticulocyte count 1% .Peripheral smear showed microcytic normochromic with few macrocytes, polychromatophils and neucleated RBCs, WBC showed myeloid shift to left. Differential counts Neutrophils 36% lymphocytes 35% eosinophils 04% monocytes 01% band forms 10% metamyelocytes 06% myelocytes 06% blasts 02%. Platelets showed moderate thrombocytopenia. Abdominal ultrasound was performed which showed fatty changes in liver and otherwise normal. He was transfused with 2 units of fresh compatible blood and again re-evaluated for severe anemia. His iron was 320 micrograms/dl (ref ranges 59-158 UG/dl).serum ferritin was 2000ng/ml (ref range 22-322 ng/ml).

Bone marrow aspirate was obtained from right posterior iliac crest, which showed marked prominence of round cells, scattered singly and also seen in small clusters at places. The cells were intermediate size around 2 to 2 1/2 the size of a small mature lymphocyte and has scant to moderate amount of cytoplasm. Some cells showed cytoplasmic vacuolations, no granules or aeur rods seen. Nucleus was round with evenly distributed chromatin and some show small neucleolus.normal hematopoietic precursors were markedly reduced.

Bone marrow biopsy report: the intervening marrow replaced by sheets of round cells at foci forming rosettes. There was moderate amount of stroma seen intervening. The cells had scant cytoplasm and round hyperchromatic nuclei. Normal hemopoietic precursors were very spars.

Bone marrow biopsy feature in correlation with immune histochemistry were in favour of marrow involvement by neuroblastoma.

Immunophenotyping of bone marrow aspirate results were negative for CD45 CD33 HLA DR, CD 19, CD22, CD10, CD2, CD4, CD7, CD13, CD33, and CD117. These features favour marrow involvement by round cell tumour.

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He underwent MRI abdomen which showed a heterogeneously enhancing lesion with areas of haemorrhage and necrosis measuring 8.1*4.8*7.9 cm encasing aorta, celiac axis and left renal artery not made out separately from left adrenal. Hepatomegaly with patchy areas of altered signal intensity and enhancement. A diagnostic possibility of ganglioneuroblastoma/neuroblastoma with metastatic deposits in liver was considered.

Whole body PET-CT scan was suggestive of metabolic active left Para aortic mass probably lymph nodal with hepatic and marrow involvement. However as the left adrenal was not seen separately a primary adrenal lesion was considered in the differential diagnosis.

Seeing to this a diagnosis of neuroblastoma stage 4 PET CT SCAN was done which showed metabolically active disease and was further subjected for chemotherapy. Now the patient became asymptomatic and was adapted for routine activity. After 6 months he underwent a whole body PET-CT scan and was compared with the previous PET-CT scan, now the PET-CT was negative for metabolically active disease

Discussion

Here we report a case of neuroblastoma in adult which is rare to occur. Neuroblastoma is a type of cancer that most often affects children. Neuroblastoma occurs when immature nerve cells called neuroblasts become abnormal and multiply uncontrollably to form a tumor. Most commonly, the tumor originates in the nerve tissue of the adrenal gland located above each kidney. Other common sites for tumors to form include the nerve tissue in the abdomen, chest, neck, or pelvis. Neuroblastoma can spread (metastasize) to other parts of the body such as the bones, liver, or skin. There was a steady decline in the incidence of adult neuroblastoma from 0.47 cases per million per year in 1973–1977 to 0.12 cases per million per year in 1998–2002 [7]. Neuroblastoma is a rare tumour, which is seen more in children rather than adults. A large percentage of neuroblastomas undergo spontaneous regression and this could possibly account for the scarcity of its presence in adult population [8]. Clinical data suggests that >40% of neuroblastomas arise from within the adrenal glands. Symptoms associated with the diagnosis are usually unrelated to the catecholamine imbalance. However on rare occasions compression of renal artery may lead to hypertension as a presenting manifestation [9]. The most common symptoms are hard asymptomatic mass in abdomen or bone pain resulting from metastatic spread. Initially we evaluated with possibility of pheochromocytoma seeing to the labile presentation of hypertension in young .In about 90% of cases of neuroblastoma, elevated levels of catecholamine or its metabolites are found in the urine or blood, which is not seen here [10]. During his second visit, symptoms and signs became more prominent with profound anemia and progressive weight loss. Anemia in neuroblastoma patients may result not only from bone marrow involvement, but also from bleeding into a tumor mass or from the haemolysis accompanying a consumption coagulopathy [11]. Bone marrow was diagnostic of neuroblastoma stage 4, serum ferritin and total iron values were significantly high suggestive of active tumour and possibility of neuroblastoma was considered. Serum ferritin level could be used as indicator of disease activity and as a guide to therapy [12]. Immunohistochemistry and immunophenotyping supported the diagnosis. Abdominal MRI scan showed a heterogeneously enhancing lesion with areas of haemorrhage and necrosis measuring 8.1*4.8*7.9 cm encasing aorta, celiac axis and left renal artery not made out separately from left adrenal. Hepatomegaly with patchy areas of altered signal intensity and enhancement. Further this patient underwent chemotherapy. PET CT was performed prior to chemotherapy and after chemotherapy.

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