Retroperitoneal Ganglioneuroblastoma in pediatric patient.

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Abstract Ganglioneuroblastoma is an uncommon tumour of the sympathetic nervous system that can originate anywhere along it. It mainly occurs in the pediatric age group with an annual incidence of 7.6 per 1,000,000 population in the USA. Ganglioneuroblastoma considered as third most common malignancy in children after leukemia and brain tumor it is also the most common solid extracranial tumor in children according to the Surveillance, Epidemiology and End Results (SEER) Registry. We report a case of 3 year and five months old girl who was admitted through the OPD for further workup for having a complaint of chronic diarrhea, failure to thrive and working diagnosis of celiac disease. Upon physical examination, she generally looks well, thin and sleeping comfortably. Weight: 12.95 Kg (2nd percentile). Height: 95.6 cm (3rd percentile). Investigations revealed that ASCA, ANCA and Celiac profile (tissue transglutaminase) were Normal. Stool electrolytes were going with secretary type of diarrhea and she was having persistent metabolic acidosis and hypokalemia raising the possibility of carcinoid tumor. Ultrasound (US), computed tomography (CT) and magnetic resonance imaging (MRI) for abdomen showed a mass in the region head of the pancreas. Laparotomy was done and tumor had been removed. Histopathology report of a removed mass demonstrated a tumor with calcifications limited to adrenal gland with microscopic foci of neuroblastic components were representing 1-5% of the whole tumor. In our literature, we have gone through different presenting complaints of the reported cases, from neck swelling, flank pain, to respiratory distress. What makes up the rarity of our case is the chronic diarrhea as the main complaint of our 3-year-old child. Therefore the rarity of mediastinal or abdominal ganglioneuroblastoms doesn't make it out of important differential diagnoses for such a vague symptom.

Key words: ganglioneuroblastoma; pediatrics; chronic; diarrhea

Introduction:

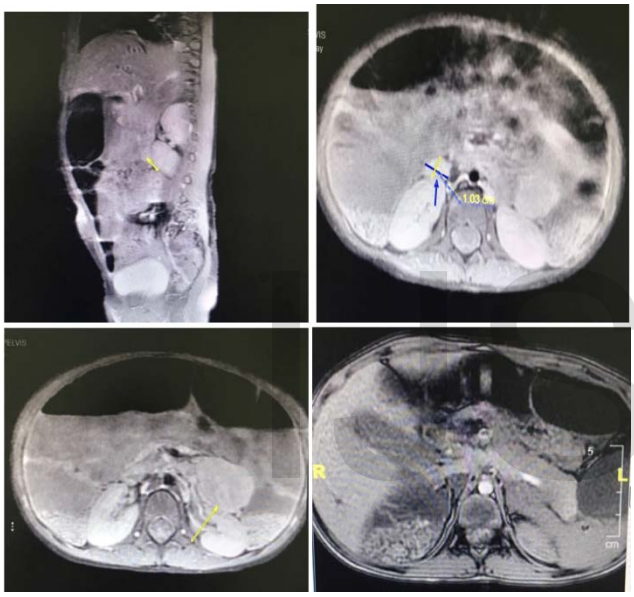
Ganglioneuroblastoma is an uncommon tumor of the sympathetic nervous system that can originate anywhere along it [1]. It mainly occurs in the pediatric age group with an annual incidence of 7.6 per 1,000,000 population in the USA [1]. Ganglioneuroblastoma considered as third most common malignancy in children after leukemia and brain tumor it is also the most common solid extracranial tumor in children according to the Surveillance, Epidemiology and End Results (SEER) Registry [2].

Case presentation:

A 3 year and five months old girl was admitted through the OPD for further workup for having a complaint of chronic diarrhea, failure to thrive and working diagnosis of celiac disease. She was seen twice in the OPD and then she admitted in the gastroenterology ward for almost one month. In her past history, she had her initial work up in other hospital and it was going with celiac disease. On physical examination: she generally looks well, thin and sleeping comfortably. Weight: 12.95 Kg (2nd percentile). Height: 95.6 cm (3rd percentile). BMI: 14.2. Tem: 36.5 c. Pulse: 107 BP: 117/60 mmHg. RR: 28. O2 sat: 99%. No abnormalities detected upon systemic examinations. ASCA, ANCA and Celiac profile (tissue transglutaminase) were Normal..

At the gastroenterology ward, she had the complaint of electrolyte imbalance with hypokalemia, persistent and metabolic acidosis which was corrected with IV fluid and oral supplement of sodium bicarbonate and potassium chloride. Stool electrolytes were going with secretary type of diarrhea and she was having persistent metabolic acidosis and hypokalemia raising the possibility of carcinoid tumor. We did an ultrasound (US), computed tomography (CT) and magnetic resonance imaging (MRI) for abdomen which showed the mass in the region head of the pancreas. Laparotomy was done on May 25th, 2017 and this tumor about the size of 3-4 cm was removed. It was retroperitoneal not attached to any organ. She remained well after the procedure and started to show improvement in her diarrhea. Upon improvement, IV fluid was discontinued and oral fluid started.

MRI Images & Findings:



A well-defined left retroperitoneal soft tissue enhancing mass lesion related to the tail of the pancreas is demonstrated. It measures 4 x 3.2cm in maximum axial dimensions and at least 5.5 cm in the longitudinal dimensions. There is a possible pre-caval lymph node measures 1.5 cm.

Pathology Report:

A tumor of 8.0 cm and 55.0 g with calcifications, limited to the adrenal gland. Microscopic foci of neuroblastic components were representing 1-5% of the whole tumor.

Discussion:

The International Neuroblastoma Pathology Classification (the Shimada system) classifies\grades peripheral neuroblastic tumors into neuroblastoma, ganglioneuroblastoma intermixed, ganglioneuroma, and ganglioneuroblastoma nodular, according to histopathological balance between primitive neuroblasts, maturing neuroblasts, and ganglion cells (neural-type cells) and Schwannian-blasts and mature Schwann cells (Schwann-type cells).[1] Ganglioneuroblastoma composed histologically of neuroblasts with mature ganglion cells; for which it fails intermediate between the most malignant neuroblastoma and the most benign ganglioneuromas.[2]

Vast majority of ganglioneuroblastoma cases are discovered before 10 years of age. [1] with only three cases of newborn found in literature. [2] It equally reported for both genders. [1] Ganglioneuroblastoma is commonly raised from adrenal medulla, extra-adrenal retroprotenium and posterior mediastinum. [1][2]

Ganglioneuroblastoma is considered a rare incidental finding for adult too. In a study included 111 adults with adrenal incidemtoma, only one found out to have a ganglioneuroblastoma. [3] The presentation of ganglioneuroblastoma either related to mass compression\pressure effects or symptoms of metastasis. [2] In our literature, we have gone through different presenting complaints of the reported cases, from neck swelling, flank pain, to respiratory distress. What makes up the rarity of our case is the chronic diarrhea as the main complaint of our 3-year-old child.

The most common sites for metastasis are bone (Hutchinson's syndrome), bone marrow, liver (Pepper syndrome) and skin ('blueberry muffin' syndrome) in patients less than one year. [2] [1]

The ganglioneuroblastoma should be first differentiated from rhabdomysosarcomam which the most common soft tissue sarcoma in pediatric age group, mostly detected in head or neck, and Wilms tumor if kidney concern. [1] The standard imaging for evaluation of ganglioneuroblastoma is computed tomography (CT) which gives from where the tumor is originating and reaching which extent plus vascular engorgement, though the definitive diagnosis is made by histology examination after fine needle aspiration or excision biopsy. [2][1] Then staging is further done

according to International Neuroblastoma Stuging study. [1] In one study, the 2 year-old patient had Horner syndrome postoperatively. And 3 patients have been reported to developed nerve paresis after surgery of cervical ganglioneuroblastoma. [4]

The mainstay of management of such tumor is surgical removal and chemotherapy post-operatively if the systematic metastasis is there. Radiotherapy and bone marrow transplant depending on the stage. [2][4] The prognosis in children with ganglioneuroblastomas originated in mediastinum is of better prognosis than those of may originate in abdomen. [2]

Conclusion\Acknowledgment:

In publishing this case, we aim to show how different and un-relevant presentations of ganglioneuroblastoma in pediatric population can be faced. The rarity of mediastinal or abdominal ganglioneuroblastoms doesn't make it out of important differential diagnoses for such a vague symptom.

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