CASE REPORT

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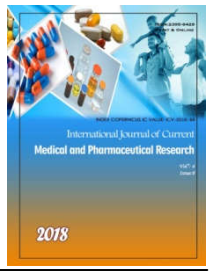

CHILD WITH STIFF GAIT: AN UNUSUAL PRESENTATION OF RARE CHILDHOOD GANGLIONEUROMA OF POSTERIOR MEDIASTINUM

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ABSTRACT

Ganglioneuroma, a rare benign tumour representative of Peripheral Neuroblastic Tumour(PNT) usually affect older children (1,2). It is predominantly localised in thoracic region but may occur at any site like intra-abdominal or retropharyngeal spaces along the occurrences of sympathetic ganglion. It may arise *de novo* or from a neuroblastoma either spontaneously or after chemotherapy. (3) The prognosis is excellent, as the surgical excision is often curative. Chemotherapy and radiotherapy have little role in its management. Here we present a young child of 5years and 3 months presented with posterior mediastinal Ganglioneuroma without any respiratory symptoms but with musculoskeletal problems like lower back pain and stiff gait.

INTRODUCTION

A 5year 3months old male patient, born out of non-consanguineous marriage, was brought to the hospital with calf muscle pain in both lower limbs along with pain and stiffness at lower back region with stiff gait. Patient was apparently well 3 weeks before, after that the child developed insidious onset and gradually progressive pain in both lower limbs mainly localising to calf muscle. Within few days he also complained of dull aching deep seated pain at lower back region without any radiation from back to lower limbs. In the course of illness, he also developed stiffness of lower back and hip region so much so that his normal gait and activity was restricted. Over the next few days the nature of pain changed from dull aching to sharp stabbing and episodic in nature involving the same lower back region and occurring 5-6 times a day. There was no involvement of other big joints and small joints, no redness or pain in eye. Patient developed low grade fever associated with mild to moderate evening rise of temperature, intermittent, not associated with chill and rigor. There was no history of associated rash, cough and cold, haemoptysis, abdominal distension, burning sensation during micturition, any alteration of sensorium or abnormal movement. The past medical history and family history was non- contributory to present illness. There was no contact history of tuberculosis and the immunisation status was as per age. The bladder and bowel habit was normal.

On examination patient was alert, conscious and co-operative, normotensive with normal cardiovascular and respiratory parameters. General survey was largely normal except for significant pallor for which he would require one unit of packed cell transfusion. Musculoskeletal examination including Paediatricgait, Arms, Legs, Spine (pGALS) examination showed definite restriction of normal forward flexion of thoraco-lumbar spine along with exaggerated lumbar lordosis. The sacro-Iliac and hip joint movement restriction and tenderness over sacro-iliac joint were also noted. There was no kypho-scoliosis and other joint examination was normal. On hip flexion beyond 50 degrees the pain used to increase. There was no hepato-splenomegaly or lymphadenopathy, no significant skin lesion, no other major organ involvement. Eye examination for uveitis was negative. Neurological examination showed normal higher neurogical function with preserved superficial and deep tendon reflexes, sensory and motor findings. Cranial nerve examination was normal and Planter response was flexor.

Routine test revealed anaemia(Haemoglobin- 6.8gm/dl), normocytic normochromic picture with elevated Erythrocyte Sedimentation Rate(ESR-78mm/1st hour).Liver Function Test, Renal Function Test, Electrolytes were within normal range. C-Reactive Protein (CRP-44mg/dl) was positive. Investigations for rheumatological profile were normal except for elevated Anti-StreptolysinO titre(ASO>200 Todd unit). Antinuclear Antibody(ANA), Antineutrophilic cytoplasmic antibody(ANCA), Rheumatoid factor, Anti- cyclic citrullinated peptide, Human Leucocyte Antigen B27 were negative. C3 value-2.31gm/l (ref 0.9-1.8), C4 value 0.59gm/l (ref 0.1-0.4) were mildly raised.

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Chest X-Ray showed a well- defined moderately large posterior mediastinal mass which was further confirmed by Contrast Enhanced Computed Tomography (CECT)of chest as "Lobulated mass with calcification seen adjacent to aortic knuckle extending to left paravertebral area with mild right pleural thickening". We excluded TB as Gastric Lavage for CBNAAT of tubercular bacilli was negative, However Mantoux test was positive at 10mm. Magnetic Resonance Imaging of dorso-lumbar spine, T2 weighted image revealed "large paravertebral soft tissue lesion involving left lung upper zone which is medially extending in spinal canal and causing pressure effect on upper dorsal cord which is showing post compressive signal changes; features suggestive of neurogenic/neoplastic mass". Spinal canal was normal and MRI of sacro-Iliac joint showed signal changes.

The Alfafeto Protein (AFP) was <1.0 ng/ml, serum Beta Human Chorionic Gonadotrophin (bhCG) was 3.5(<5.0mIU/ml). The urinary Vanillylmandelic Acid (VMA) was raised further corroborating neurogenic mass. Fine Needle Aspiration Cytology of the mass was non-specific for which CT guided biopsy done which showed "A lesion composed of interlacing fascicles of spindle shaped cells having elongated nuclei and tapering ends lying in a collagenised stroma, scattered ganglion cells are present. The tumour cells do not show significant nuclear pleomorphism or mitotic activity".

All the above findings were suggestive of Childhood Ganglioneuroma". We consulted for Cardio-thoracic-vascular surgery and Neurosurgical opinion for further treatment and feasibility for surgical intervention.

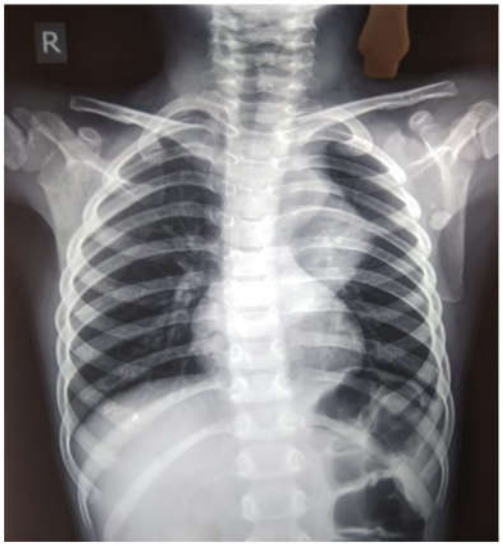

Fig 1 Chest X-ray showing mediastinal mass

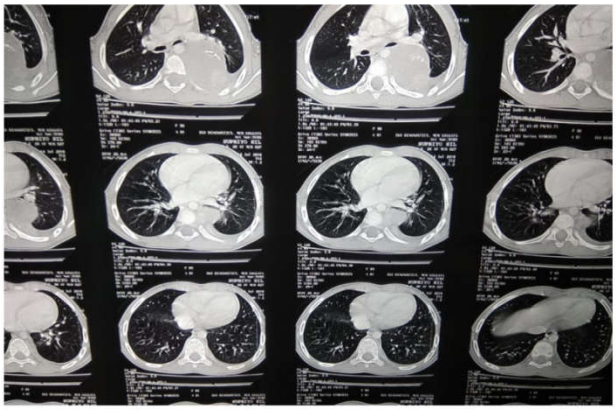

Fig 2 Contrast Enhanced CT chest showing mediastinal mass

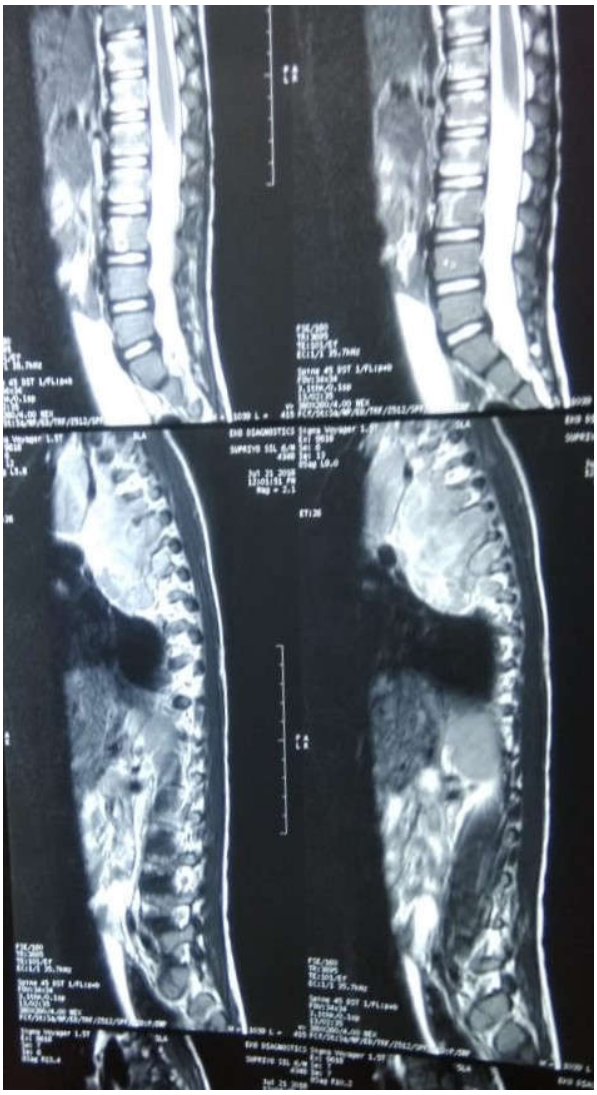

Fig 3 Magnetic resonance image of thoracolumbar spine shows posterior mediastinal mass encroaching spinal canal.

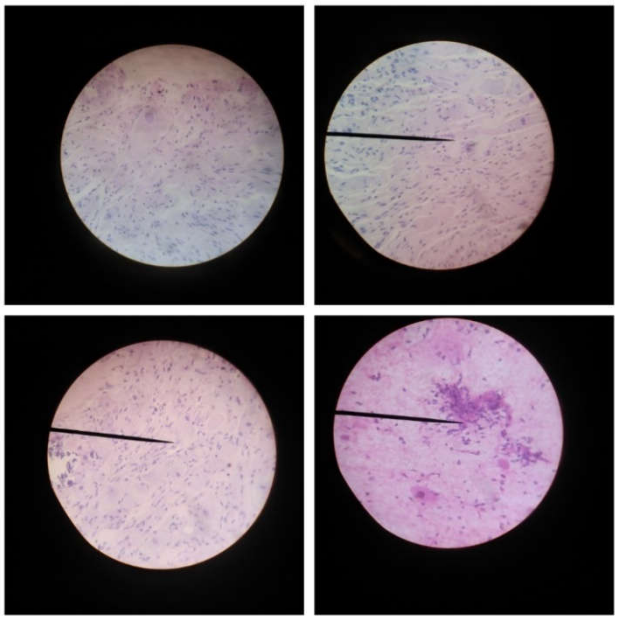

Fig 4 Histopathology showing interlacing fascicles of spindle shaped cells having elongated nuclei and tapering ends lying in a collagenised stroma, scattered ganglion cells without any features of neoplasia.

DISCUSSION

Ganglioneuroma(GN) is a benign neurogenic tumor. These tumors are originating from neuroepithelium along sympathetic ganglia. It is one of the three spectrum of Peripheral Neuroblastic Tumour(PNT), the other two being Neuroblastoma (NB) and Ganglioneuroblastoma (GNB).(1) Main localization is the posterior mediastinum in children older than 10 years followed by the retro peritoneum and cervical region.(4). Ganglioneuromas are asymptomatic in most cases and symptoms are usually caused by mass effects. Imaging studies are showing a lobulated and calcified mass, and several differential diagnoses have to be considered, i.e. neurofibroma or chordoma. (5) Ganglioneuroma can arise *de novo* or as a result of maturation of a neuroblastoma / Ganglioneuroblastoma, either spontaneously or secondary to treatment of a neuroblastoma. An association with malignant neuroblastoma is rarely observed and it still remains a topic of discussion.(4) Ganglioneuroma, according to International Neuroblastoma Pathology Classification(INPC criteria), is composed predominantly of ganglioneuromatous stroma with a minor component of scattered, evenly or unevenly distributed collections of differentiating neuroblasts and / or maturing / mature ganglion cells.(2) Hence, a complete excision and proper histopathological examination of the entire specimen is required, to rule out foci of immature neuroblasts as has been carried out in the index case.(1) Late malignant transformation of a benign lesion to neuroblastoma, thoughrare, has also been reported.(6) In contrast to neuroblastoma, secretion of catecholamine is an uncommon finding in ganglioneuromas. 80% of neuroblastoma are producing elevated levels of Vanillylmandelic Acid(VMA) and Homovanilic Acid (HMA) and urine tests are used as screening method.(4) In our case there was elevated VMA level suggesting increased catecholamine activity of Ganglioneuroma as well which will require further follow up.

Posterior mediastinal mass in our case was an incidental finding. The differential diagnosis of posterior mediastinal mass is mainly neurogenic mass like Nerve sheath tumours (Schwannoma, neurofibroma, malignant peripheral nerve sheath tumour), Parasympathetic ganglionic tumour (paraganglioma, chemodectoma, pheochromocytoma), Sympathetic ganglionic tumour (ganglionuroma, ganglioneuroblastoma, neuroblastoma).In the study by Heimburger *et al* 24% were the posterior mediastinal tumours of all the mediastinal tumours of which 82 per cent were of neurogenic origin and of these 80 per cent were from sympathetic chain.(7)]. Ganglioneuroma was seen in only 11 patients out of 186 mediastinal masses in the series by Rubush *et al* (8), and 2 out of 42 cases in the series by Heimburger(7). The ganglioneuroma cases mostly present after the age of 10 years, only 9 patients of 88 cases were between 5-9 years (9).

The basis of the clinical presentation here is compression effect of the mass encroaching inside spinal canal causing root pain and muscle spasm at thoracolumbar region with referred pain at the hip joint and sacroiliac joint.

The definitive treatment of choice for GN is usually complete surgical resection. Most tumours can be excised totally. If the tumour is catecholamine secreting, the acute and chronic effects of increased plasma catecholamines should be reversed prior to the surgical excision of the tumour. Combined α-and β-adrenergic blockades are required preoperatively to control high blood pressure and to prevent intraoperative hypertensive crises. Laparoscopic surgery for abdominal GNs may be a better substitute for traditional open surgery due to it is minimal invasive procedure, especially in tumours with a smaller than 6 cm in diameter. Overall, patients with GN have a favourable prognosis. The recurrence rate is near zero, and postoperative complications are rare. However, life-long clinical and biochemical follow-up patients with hormone-secreting GNs, transformation to neuroblastoma,adrenal composite pheochromocytoma-GN and metastatic disease is essential.(10)

Key message: Ganglioneuroma should be one of the differential diagnosis when patient present with rheumatological manifestations as well as lower back pain and muscle stiffness.

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