Case Report

GANGLIONEUROMA OF MEDIASTINUM AND ADRENAL GLANDA CASE REPORT

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ABSTRACT

Introduction: *Ganglioneuroma is a rare benign nervous tumor arising from sympathetic ganglion which commonly occurs at posterior mediastinum, retroperitoneum and adrenal gland.*

Case**:** *We report a 6 year-old boy who presented with abdomial pain. Compute tomography scan and magnetic resonance imaging (MRI) showed one tumor at his right posterior mediastinum area measuring 8.3x6.3x5.3cm, and the other tumor at his right adrenal gland measuring 3.9x3.4x3.2 cm. A biopsy showed ganglioneuroma, and complete resection of both tumors. Post operation biopsy confirm definitive diagnosis as ganglioneuroma. The patient is healthy from surgery.*

Conclusion**:** *Ganglioneuroma is benign. The treatment is complete surgical resection. Prognosis after surgical resection seems to be so good, without any recurrences or need for adjuvant therapy.*

I. INTRODUCTION

Ganglioneuroma is a rare and benign neurogenic tumor arising from central or peripheral parts of the autonomic system [1,2]. Gangioneuroma most commonly occurs in the posterior mediastinum, retroperitoneum and adrenal gland. It rarely appears in the cervical, head regions [3]. It is commonly asymptomatic and diagnosed by chance; however, it could lead to compression sydrome due to tumor size and rarely to systemic symptoms [4,5]. The results of computed tomography (CT) scan, magnetic resonance imaging (MRI) and ultrasound could suggest gangioneuroma. However, histopathologic examination is the only tool to confirm ganglioneuroma and to differentiate it from other neural crest tumor [6]. The treatment is complete surgical resection. The treatment is complete surgical resection [6]. Most ganglioneuroma is noncancerous, so the prognosis is commonly good.

II. CASE REPORT

Our patient now aged 6 years old, was born on Danang city, Vietnam. The child felt abdominal pain, and was done ultrasound and found incidentally one tumor in his right adrenal gland. Then he was done CT scan and whole body MRI. The results

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revealed one mass measuring 8.3x 6.3 x 5.3 mm at the posterior mediastinum with some calcifications inside (Figure. 1) and one other mass measuring 39 x 34 x 32 mm at the right adrenal gland (figure. 2). Both masses are mainly hypodense on arterial imaging and heterogenous on delayed phase images.

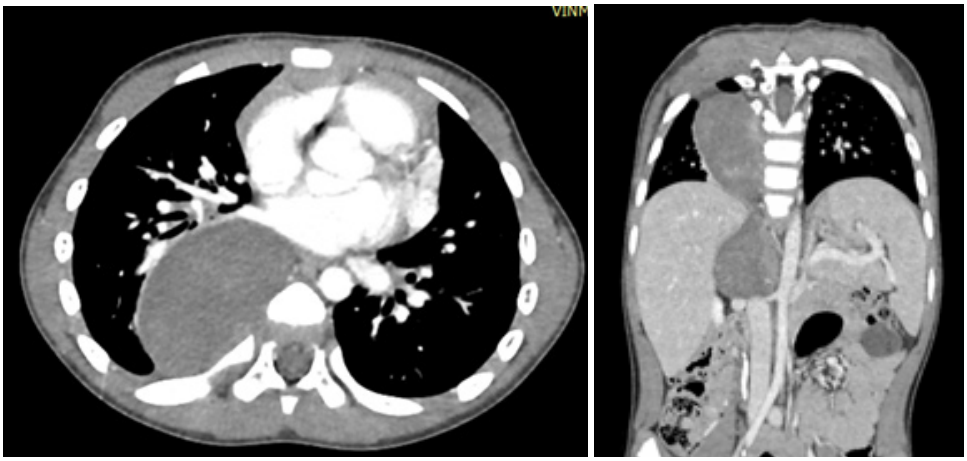

Figure. 1: Computed tomography scan of thorax showing mass in the right posterior mediastinum region

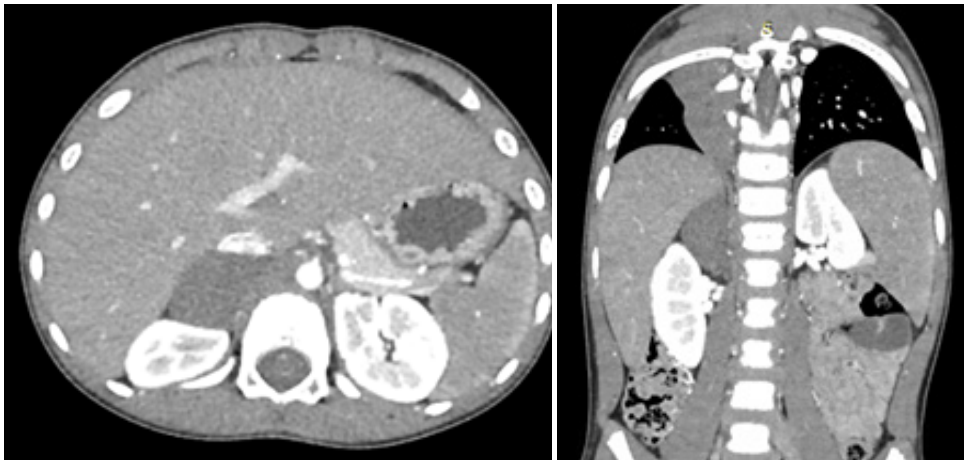

Figure. 2: Computed tomography scan of abdomen showing mass in the right adrenal gland region

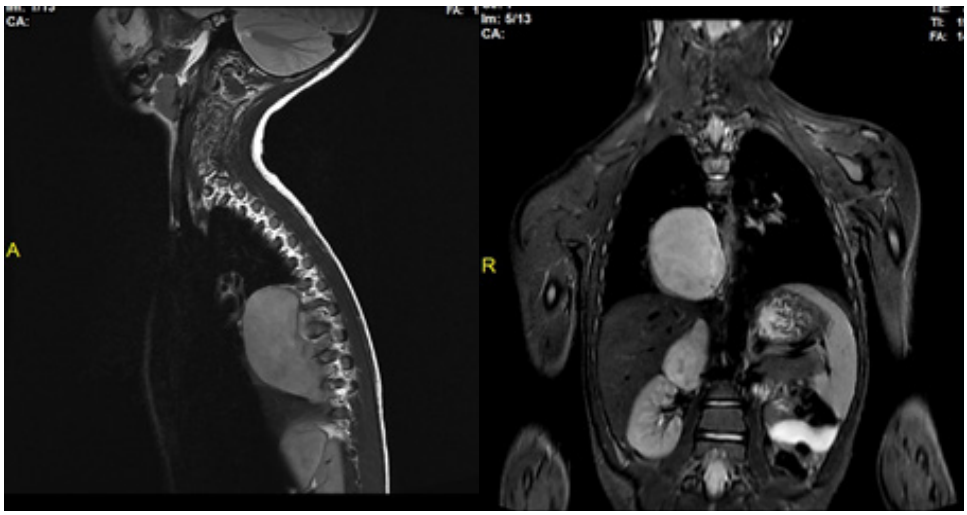

Figure. 3. Magnetic resonance imaging showed the mass in both posterior mediastinum and adrenal gland

The child was transferred to Hue Pediatric Center. We examined him carefully: he was alert without hypertension. He did not have respiratory distress and abdominal pain. He was done bone scan, full blood count, liver and kidney function, electroylyte test, LDH, acid uric. The results showed LDH slight elevated (285 u/l). The other tests were normal. A needle biopsy of his thoracic mass showed mature ganglion cells and Schwann cells.

After multidisciplinary team discussion, as well as receiving consultations from experts, surgical resection was recommended. Both masses were removed completely. Pathology examination after a complete resection was consistent with the diagnosis of ganglioneuroma. The pathological hematoxylin-eosin stains disclosed mature ganglion cells with eosinophilic cytoplasm with distinct cell borders, single eccentric nucleus, prominent nucleolus and Schwann cells.

After surgery, the child recovered uneventfully. He is healthy now and the ultrasound does not show any residual or recurrent mass.

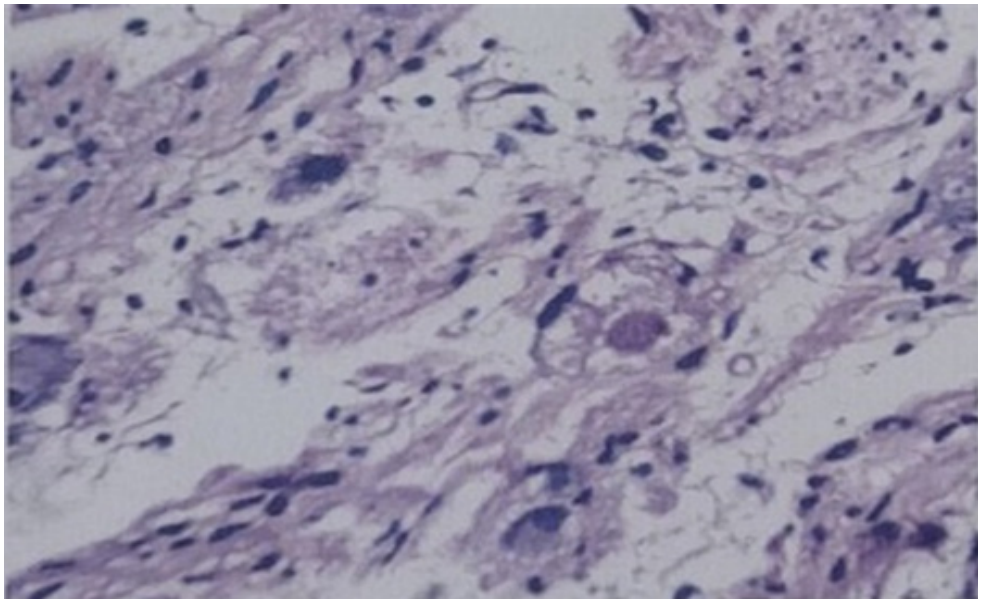

Figure. 3. Histopathology of ganglioneuroma. HE stain showing mature ganglion cells and Schwann cells.

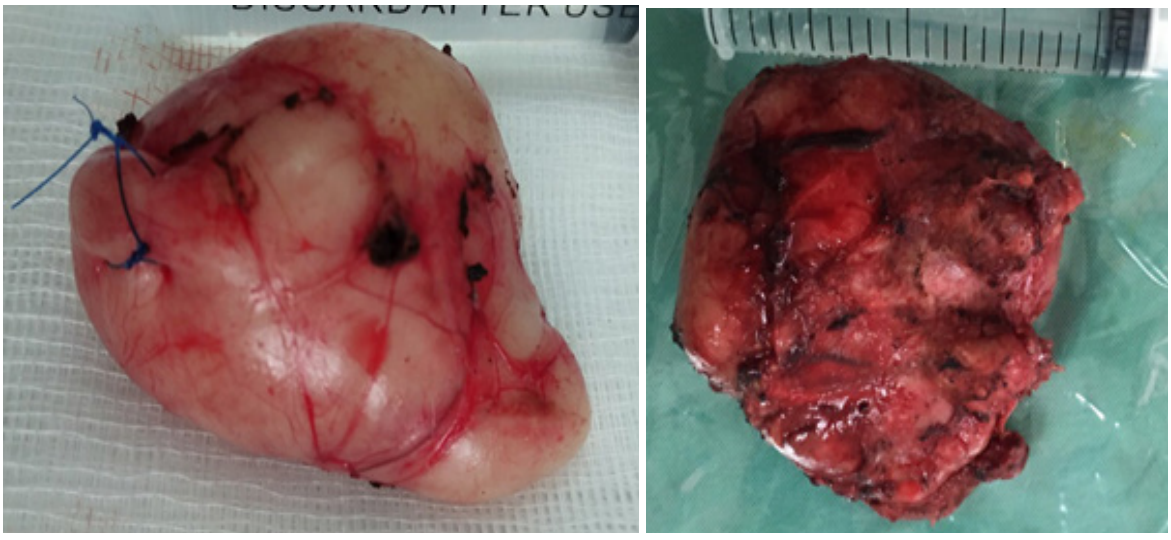

Figure. 4. Excised specimens of posterior mediastinal mass and adrenal mass

III. DISCUSSION

Ganglioneuroma arises from cells of the neural crest including sympathetic ganglia and adrenal glands. It generally occurs in thoracic (41.5%), retroperitoneal region (37.5%) and adrenal gland (21%). Other sites such as the head and neck can also be affected (8%). It affects children and young people with three-fifth developed before age 20. Female are more prone to be affected than males [4,6-8].

Ganglioneuroma is a benign and slow-growing tumor, mosty hormonally silent. It is typically asymptomatic, and is found incidentally when being examined or treated for another condition. Any symptoms will depend upon the tumor's location and the nearby organs affected [6,7,9].

Imaging studies such as ultrasonography, CT and MRI could suggest diagnosis. A well-defined mass with homoechogenicity is often revealed by ultrasonography and CT. Ganglioneuroma appears as an encapsulated mass that is enhanced slightly by contrast medium. Calcification might be found within the tumor, as in our patient. MRI shows a homogeneous mass with signal intensity less than that of liver on T1- weighted images, and as a heterogeneous mass with predominant signal intensity greater than that of liver on T2-weighted images [10,11]. However, histopathologic examination is the only tool to confirm ganglioneuroma and to differentiate it from other neural crest tumor. It is composed of ganglion cells, Schwann cells and fibrous tissue.

The most significant differential diagnosis of ganglioneuroma is neuroblastoma. Increased level of urinary noradrenaline, dopamine, HVA, and VMA are frequently encountered in neuroblastoma, while the level of urinary cathecolamine, HVA, and VMA are usually normal in ganglioneuroma. In our case, we could not measure VMA, HVA. However, the pathology confirmed the diagnosis of ganglioneuroma.

Although ganglioneuroma is benign, surgical removal is the mainstay of treatment [12], as it could cause pain or compression symptoms, could be locally aggressive and can lead to cord compression. In addition, pathology of the resected specimen can confirm the diagnosis [5]. In general, prognosis after surgery seems to be good, without any recurrences or need for adjuvant therapy.

IV. CONCLUSION

Ganglioneuroma is uncommon and benign tumor arising from neural crest cells. The lesions are often found incidentally and tend to be asymptomatic. CT scan and MRI could suggest diagnosis. However, pathology is a golden criteria to diagnosis. Complete surgical excision is the gold standard for the treatment of posterior and adrenal ganglioneuroma. Post surgery, there is no need for adjuvant treatment and the overall prognosis is excellent.

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