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Case Report

Mediastinal ganglioneuroma with osseous invasion simulating malignant transformation of osteochondroma on CT imaging☆

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ABSTRACT

Ganglioneuromas (GN) are rare, mature tumors that arise in the posterior mediastinum or retroperitoneum from neural crest cells and present as slow growing masses in the pediatric population. While they are often found incidentally in unrelated diagnostic workup, they can become symptomatic due to their size and location. They typically demonstrate the nonspecific appearance of a solid mass without invasive or destructive features across different modalities. Such features are normally indicative of more aggressive neoplasms from similar cellular ancestry or an entirely different lineage. Here we present a case of mediastinal GN that on imaging was initially suggestive of an osteochondroma with malignant degeneration based on the presence of an exostosis associated with a large solid mass. Final pathology, however, revealed GN with involvement of the adjacent bone. While the final diagnosis was benign, it is important to recognize this pattern of exostosis with solid mass, especially since the overall survival rate of sarcomata is much worse than that of a classic GN.

Introduction

Ganglioneuromas (GN) are benign tumors that originate from neural crest tissues of the sympathetic ganglia most commonly in the mediastinum or retroperitoneum [1]. Patients with these tumors are often asymptomatic and are incidentally detected on imaging for other purposes in late childhood [2]. Although benign and often asymptomatic, intrathoracic GN may grow aggressively compressing vital structures such as the spinal cord and tracheobronchial tree leading to neurologic and respiratory symptoms [3–5]. The CT appearance of a

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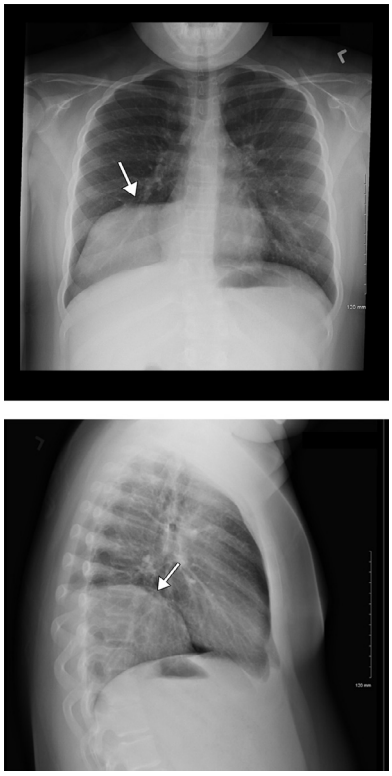

Fig. 1 – A. PA radiograph of the chest demonstrating soft tissue mass in the lower right hemithorax (arrow). B. Lateral view localizes the mass to the posterior mediastinum (arrow). Osseous excrescence and calcifications are not evident on radiograph.

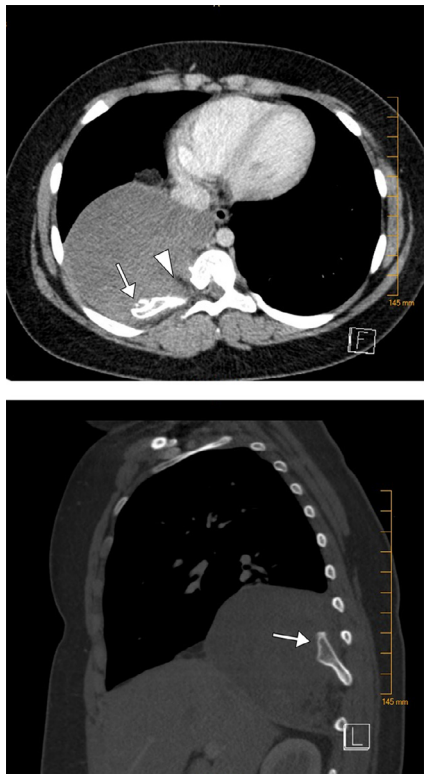

Fig. 2 – A. Axial CT image of the chest at the level of the 10th rib demonstrating osseous excrescence in the right hemithorax with associated soft tissue mass, coarse calcifications (arrow) and expansion of the neural foramen (arrowhead). B. Sagittal reconstruction demonstrating the full extent of the osseous excrescence with continuous medullary cavity arising from the 10th rib (arrow).

GN is usually nonspecific, however the differential diagnosis tends to be limited to other neuroblastic tumors given their characteristic location. Here we present an interesting case of a symptomatic mediastinal GN in a young female whose initial imaging demonstrated an unusual exostosis suggestive of osteochondroma with malignant degeneration.

Case report

A 12-year-old female presented to her primary care provider after experiencing chest pain, chest tightness, and shortness of breath for the previous 2-3 weeks. Additional symptoms included intermittent cough, nausea, right shoulder pain, and back pain. Physical exam was unremarkable except for obesity and decreased lung sounds upon auscultation in the right middle and lower lobes. An initial chest x-ray was performed and showed a large right lower lobe mass (Fig. 1). A subsequent chest CT with contrast was performed confirming a 10 × 9.1 × 9.5 cm heterogenous soft tissue mass in the posterior right hemithorax associated with a 2.8 cm pedunculated osseous excrescence arising from the posterior aspect of the right 10th rib with a continuous medullary cavity (Fig. 2). The lesion demonstrated internal course calcifications and minimal fat density components. There was mild widening of the T10th and 11th neural foramen on the right. Based on these findings, the diagnostic impression included osteochondroma with the possibility of sarcomatous degeneration followed by neuroblastic tumors. A Pediatric Pulmonologist conducted spirometry which demonstrated borderline obstructive pattern with no change after bronchodilator administration. Complete blood count and complete metabolic panel were unremarkable. Blood screening and lack of significant constitutional symptoms were reassuring and based on imaging findings of rib changes, vertebral body changes, and foraminal involvement, GN was favored followed by osteochondroma. The patient underwent combined thoracoscopic/thoracotomy surgical exploration where a right posterior mediastinal mass was confirmed and resected along with a portion of the 10th rib. After surgery, she was admitted to the hospital for monitoring and recovery. Her hospital course was unremarkable, and she was discharged on postoperative day 2.

Final microscopic examination of the specimen showed histologic features diagnostic of GN: a spindle cell tumor composed of fascicles of bland spindle Schwann cells with area of myxoid stromal change and focal lymphohistiocytic infiltrate (Fig. 3B). Focally present within the Schwannian background were clusters of mature ganglion cells (Fig. 3C)

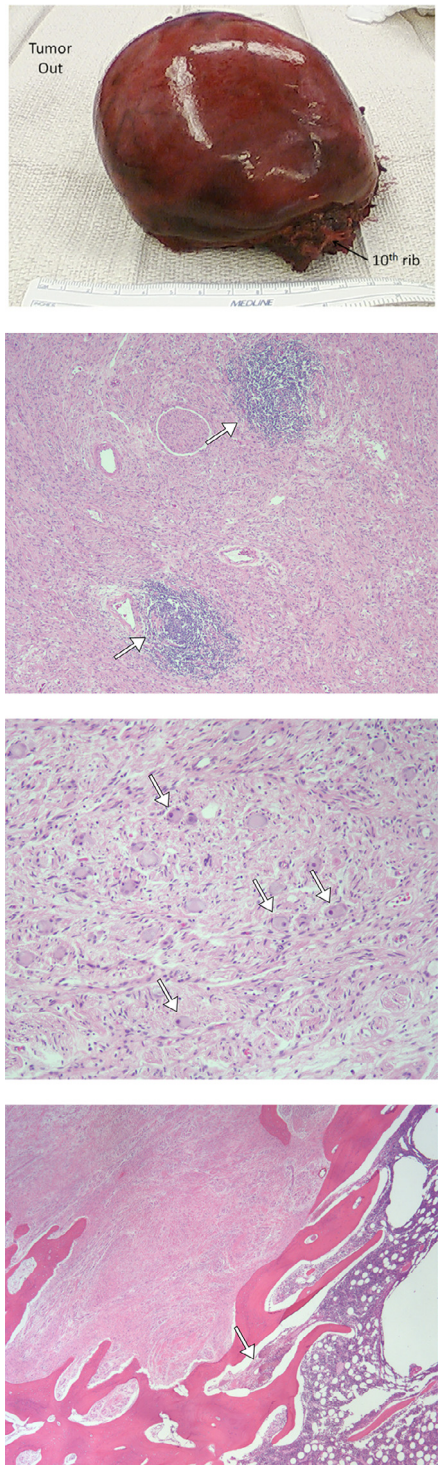

Fig. 3 – A. Macroscopic view of the specimen. Surgical specimen shows a well circumscribed tumor with a slightly lobular edge. B. Tumor composed of fascicles of bland spindle Schwann cells with focal myxoid stroma and lymphohistiocytic infiltrate (Hematoxylin & Eosin, 4X magnification). C. Clusters of ganglion cells (arrow) in the background of Schwannian stroma are seen (Hematoxylin & Eosin, 10X magnification). D. The tumor is infiltrating into bone and marrow cavity (Hematoxylin & Eosin, 4X magnification).

with eccentrically located nuclei, prominent nucleoli, dense eosinophilic granular cytoplasm, and distinct cytoplasmic membrane. There were no immature elements nor Homer Wright rosettes, cellular atypia, or mitotic figures to suggest malignancy. However, the tumor was infiltrating into bone and marrow cavity intermixing with hematopoietic cells of the marrow cavity (Fig. 3D). The tumor was present at the resection margins both macroscopically and microscopically.

Discussion

The differential diagnosis of posterior mediastinal masses in the pediatric population is usually limited to peripheral neuroblastic tumors [6]. This group of tumors arise, in varying degrees of differentiation, from the primitive neural crest cells that will eventually form the sympathetic ganglia. The term includes neuroblastoma, ganglioneuroblastoma, and GN, with the latter representing the most differentiated variant composed entirely by mature ganglion cells and other mature tissues [7].

GN represents a small fraction of all neuroblastic tumors, less than 6% in most series [7,8]. It generally occurs in the older pediatric population with median age of diagnosis ranging from 5.5 to 10 years of age. Given its cellular origin, GN is normally found in the same anatomical distribution of the sympathetic chains, with the posterior mediastinum being the most frequently affected (40%) followed by the retroperitoneum (37%) [9]. The indolent nature of the tumor allows for slow and steady growth while the patient remains asymptomatic. Most symptoms arise from compression of mediastinal structures and lung parenchyma as the tumor increases in size. As many as 40% of GN show catecholamine production and secretion of these compounds is enough to cause sympathetic activation symptoms in rare cases [9,10].

GN typically appears as a uniform hypodense mass in the posterior mediastinum with mild to moderate enhancement. Speckled or coarse calcifications are present in 40%-60% of cases [11]. Unfortunately, malignant variants of neuroblastic tumors can have an indistinguishable appearance on CT. Additional findings like necrosis, hemorrhage, vascular encasement, osseous scalloping, and foraminal expansion can be helpful to favor malignant etiology but only the presence of metastatic disease is highly suggestive [6,11]. MR imaging is the best tool to evaluate local extent and invasion of adjacent tissues and spaces. GN typically shows bright T2 signal and Low T1 signal with slow mild to moderate enhancement. While MRI was previously considered incapable of differentiating GN from malignant variants, recent studies evaluating the use of diffusion weighted imaging indicate that high apparent diffusion coefficient (ADC) values can reliably differentiate GN from neuroblastoma and ganglioneuroblastoma [12–14]. Ultimately histopathologic analysis is necessary for definitive diagnosis.

An osteochondroma is an osseous outgrowth with a cartilaginous cap that arises from bones with endochondral ossification. It is often identified as the most common tumor like lesion of the bone and typically affects the long bones of the lower extremities in close proximity to the physis [15]. They

represent benign developmental abnormalities rather than true neoplasms and most often manifest only as a small palpable mass or tenderness. Small flat bones and the axial skeleton are less frequently affected but tend to demonstrate unusual presentations (such as pneumothorax and thoracic outlet syndrome). In typical cases, plain radiography is enough to obtain a diagnosis. Other imaging modalities like CT or MR are often necessary to demonstrate the typical configuration when the lesion arises in unusual locations and to evaluate for possible complications like neurovascular compression, bursa formation and malignant degeneration.

Malignant degeneration is the most feared complication and occurs approximately in 1% of all cases, frequently in association with Hereditary Multiple Exostoses [15]. It most commonly presents as chondrosarcoma arising from the cartilaginous cap, but osteosarcoma arising from the osseous stalk is also well described. Several radiologic and clinical findings have been associated with increased likelihood of sarcoma: growth after skeletal maturity, developing lucency, cortical destruction, pain after puberty, and associated soft tissue mass or cartilaginous cap thickness above 1.5 cm. A detailed discussion of the radiologic appearance of sarcomatous masses is beyond the scope of this article, but it is worth noting that while CT and MRI is often able to differentiate CS and OS, select cases in unusual locations or with uncharacteristic mineralization can mimic other indeterminate soft tissue masses with calcification, making histopathologic examination necessary for definitive diagnosis.

Preoperative imaging in our case was challenging given the presence of an osseous outgrowth arising from the 10th rib demonstrating the typical radiologic appearance of an osteochondroma. Additionally, the presence of an associated soft tissue mass and abnormalities of the adjacent vertebral peduncles and neural foramina were highly concerning for a malignant process. Our literature review did not yield similar cases of imaging overlap involving GN and malignant degeneration of osteochondroma. The presence of GN bone invasion in the surgical pathology is also a rare finding in the current literature, however it is not clear if the bony outgrowth was present prior to tumor development or it developed as a result of tumor growth.

Conclusion

Neuroblastic tumors are overall rare neoplasms, and GN represents only a small fraction among them. Our understanding of their presentation on imaging is largely limited by their low incidence. Larger series are required to allow a more complete characterization of their appearance and to identify useful features to aid in their differentiation. In retrospect, while the findings in our case were highly suspicious for an alternative diagnosis, the demographics, clinical presentation, and classic location of the lesion should have been regarded with higher value. Additionally, the relative paucity of mineralized matrix in relation to the size of the mass can also argue against osteochondroma/chondrosarcoma. It is important to note that in some cases the alternative diagnosis may carry great morbidity and poor prognosis, which should be made clear in the final report. In challenging cases like these, additional multimodality imaging and a multidisciplinary evaluation with targeted biochemical testing could provide a greater degree of certainty prior to an invasive surgical treatment.

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