Journal of Biosciences and Medicines, 2024, 12, 30-37
https://www.scirp.org/journal/jbm
ISSN Online: 2327-509X
ISSN Print: 2327-5081

One Case of Right Posterior Mediastinum Intraneural Hemangioma Misdiagnosed as Neurilemmoma

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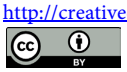
How to cite this paper: Tan, X.X., Zhang, R.F. and Luo, L.P. (2024) One Case of Right Posterior Mediastinum Intraneural Hemangioma Misdiagnosed as Neurilemmoma. *Journal of Biosciences and Medicines*, **12**, 30-37.
https://doi.org/10.4236/jbm.2024.127004

Received: May 16, 2024
Accepted: July 6, 2024
Published: July 9, 2024



Abstract

Although rare, intraneural hemangiomas should be considered in the differential diagnosis of peripheral nerve lesions. We report on a 59-year-old female patient, who was admitted to the hospital due to the discovery of bilateral breast masses for 3 months, there was no paresthesia or dyskinesia. The patient accidentally found a mass in the right upper mediastinum while completing a plain chest X-ray, initially suspected as a benign neurilemmoma on CT. Surgical resection and pathological analysis confirmed an intraneural hemangioma. Unexpectedly, the patient developed new-onset right upper limb numbness and paresthesia 3 months post-operatively, probably related to surgical nerve injury. This case underscores the importance of maintaining a broad differential for mediastinal masses, and the potential for iatrogenic neurological complications when managing these rare, yet vascular lesions.

Keywords

Intraneural Hemangioma, Mediastinal Tumors, Spinal Nerve

1. Introduction

Hemangiomas predominantly affect young individuals, with a mean age of onset around 40 years and a female predominance [1]. While these vascular lesions can occur throughout the body, mostly in the skin of the head and neck, and commonly in some organs such as the liver, they are exceptionally rare in the mediastinum, accounting for less than 0.5% of all mediastinal tumors [2]. Superficial hemangiomas appear as cutaneous discolorations or soft, compressible masses that may vary in size with postural changes. However, intraneural he-

mangiomas located in the posterior mediastinum can be easily misdiagnosed for the more common neurilemmomas, as they lack the characteristic cutaneous findings and are deeply situated. An intraneural hemangioma is a very rare, benign mesodermal lesion [3], which can involve any nerve, the lesion usually grows along the direction of the nerve bundle, and most patients will present with paresthesia or dyskinesia associated with the affected nerve [4]. This article is to emphasize the importance of maintaining broad identification for uncommon mediastinal lesions to avoid misdiagnosis and ensure appropriate treatment. We have obtained all appropriate patient consent forms.

2. Case Report

A 59-year-old woman presented with a history of recurrent bloody nipple discharge from the right breast for 4 years, ultrasound found mass in both breasts three months ago. The patient was admitted to the hospital for surgical treatment. Chest X-ray (**Figure 1**) showed the right upper mediastinum was widened and a kind of round high density mass was seen, the boundary was clear, and the lesion overlapped with the spine on the lateral radiography. Contrast-enhanced CT of the chest (**Figure 2**) showed a quasi-circular object mass, measuring 2.2 cm × 2.9 cm × 3.0 cm, on the right side of the spine at the level of the T2 - T3 vertebral body. The boundary was clear, the density was uneven, and there was no calcification within the mass. The lesion locally extended to the right intervertebral foramen, and the right intervertebral foramen was slightly enlarged at the corresponding level. There was no sign of bone destruction in the adjacent vertebral body and ribs, and the surrounding lung tissue was slightly compressed, the arterial phase of enhanced scan showed nodular enhancement at the edge of the lesion, and the venous phase enhancement was filled to the center, which was considered to be benign and likely to be a neurilemmoma. Notably, the patient remained entirely asymptomatic, denying any neurological deficits or related complaints. The patient was in good general health and denied relevant past medical history and trauma history. Laboratory studies were unremarkable.

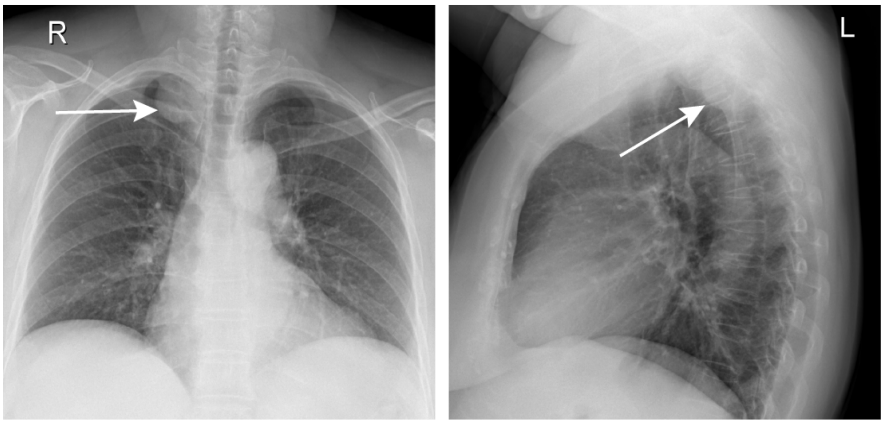


Figure 1. Chest radiography revealed a right upper mediastinal occupation (white arrow), with the lateral view demonstrating the mass (white arrow) overlapping the spine.

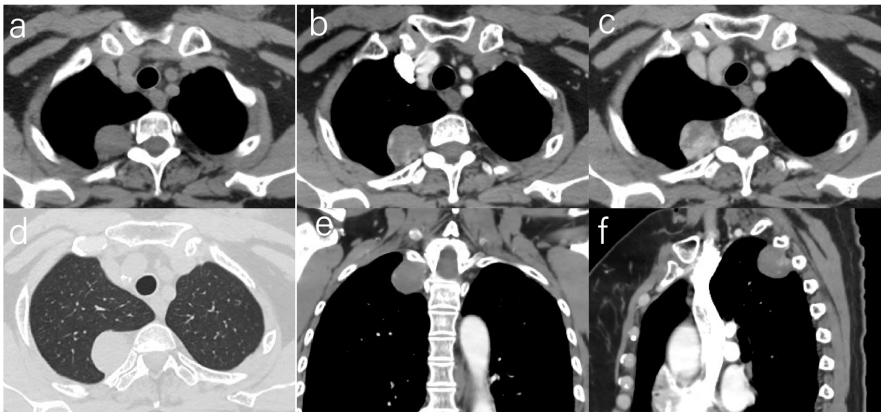
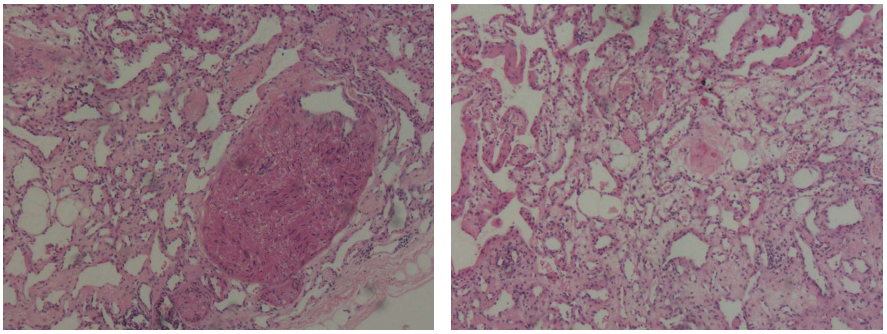


Figure 2. Chest CT plain scan mediastinal window (a) the right side of the spine showed a circular mass, connected to the pleura with a broad base, and the pleura moved in. In arterial phase axial position (b), exhibited peripheral nodular enhancement, while the venous phase (c) demonstrated progressive centripetal filling of the mass, suggestive of a vascular neoplasm. The pulmonary window (d) reveals the lesion at an obtuse angle to the chest wall, indicating that the lesion is located outside the lung. (e) and (f) represent the coronary and sagittal positions, respectively, in the arterial phase of contrast-enhanced scanning.

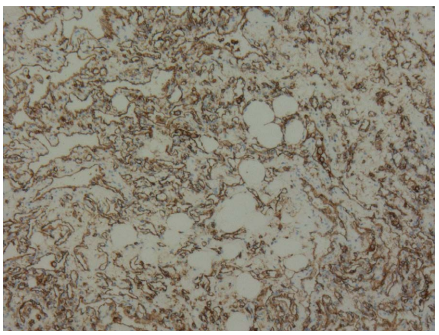
The patient underwent thoracoscopic resection of the right posterior mediastinal mass and intercostal nerve block therapy. The patient was positioned in left lateral decubitus, the incision was made into the chest at the 7th intercostal place 1cm from the right posterior axillary line and the 4th intercostal place 3 cm from the right anterior axillary line, thoracoscope was implanted and instruments were operated respectively. Intraoperatively, no fluid was found in the right thoracic cavity, a well-circumscribed, firm, gray-white tumor measuring approximately 2 × 3 × 3 cm was identified in the right posterior mediastinum, adjacent to the spine. The pleura surrounding the tumor was separated by electric hook and ultrasonic knife, the tumor was completely removed. Intraoperative frozen section analysis confirmed the lesion to be a vascular neoplasm. Following surgery, a drainage tube was inserted into the right thoracic cavity. Post-operative care included symptomatic treatment such as pain management, infection prevention, and nutritional support. On the first day post-surgery, the patient complained of pain in the operative wound and had a fever of 39˚C, which was managed symptomatically. Subsequent chest X-ray confirmed lung re-expansion and smooth drainage tube function, with 50 ml of bloody pleural effusion removed. After drain removal, the patient was advised to mobilize to prevent complications. By the 6th day, the patient was discharged in good condition, with instructions to avoid strenuous activity for a month and to attend follow-up appointments. Arranging wound care and stitches removal according to wound healing. This comprehensive approach aims to ensure optimal recovery and minimize complications for the patient's well-being.

Grossly, the resected specimen exhibited a gray-white to gray-red, firm cut surface. Microscopically, the tumor was located within the nerve, growing between the nerve fascicles and the epineurium (**Figure 3(a)**). The lesion was

composed of thin-walled vascular channels and small muscular vessels, containing erythrocytes within the lumina and lined by flat or cuboidal endothelial cells, without evidence of mitotic figures (**Figure 3(b)**). Immunohistochemical staining was positive for the endothelial markers CD31 (**Figure 3(c)**), CD34, and ERG, but negative for S100, Desmin, MelanA, and D2-40. The Ki-67 proliferative index was low, at approximately 1%. Based on these histopathological and immunohistochemical findings, the final diagnosis was confirmed as an intraneural hemangioma.



(a) (b)



(c)

Figure 3. (a) Histopathological examination revealed the intraneural location of the tumor (H & E, ×40). (b) Numerous small vascular channels lined by flat or small cubic vascular endothelial cells are demonstrated (H & E, ×40). (c) Immunohistochemistry confirmed the endothelial lineage, with positive staining for the vascular marker CD31 (CD31, ×40).

3. Discussion

When encountering posterior mediastinal masses, the common assumption is to prioritize neurogenic tumors in the differential diagnosis, and some details of the lesions are ignored, which can leads to misdiagnosis. Neurilemmomas' radiographic characteristics present as a well-defined, circular mass, it may be accompanied by calcification or cystic degeneration, the lesion grows along the long axis of the nerve bundle, the adjacent vertebral space is enlarged, generally without surrounding structures invasion, which is similar to hemangiomas, some hemangiomas can invade bones [5]. The main distinguishing point between the two is that neurilemmomas are rich in lipid Schwann cells and adi-

pocytes. The interstitial fluid of a schwannoma containing Antoni B tissue fused to form a cyst cavity, so the enhanced scanning showed significantly uneven enhancement. While intraneural hemangiomas are composed of vascular lumens of varying sizes, the lumen is separated by fatty fibrous tissue, the contrast agent enters the tumor from the blood supply vessel, then each vessel cavity is gradually filled through the fat fiber spacing, finally, all the vascular lumens in the tumor were filled with contrast agent, therefore, the enhanced scanning was progressively filled. In addition, a unique radiographic feature of intraneural hemangiomas was the virtual disappearance of the mass on repeated rotation even though it was sharply delineated on anteroposterior and posteroanterior views [6].

Ultrasound can offer real-time, dynamic assessment of the tumor's vascular distribution and its relationship to surrounding structures [7]. On ultrasound, introneural hemangiomas typically appear as enlargement and thickening of the affected nerve, with multiple anechoic tubular structures visible along the nerve's long axis, while the nerve fascicles remain continuous [8]. Some of these vascular channels may demonstrate slow, venous-pattern blood flow on Doppler evaluation, and increased flow may be observed with probe compression, whereas insufficient flow in certain lesions may indicate intratumoral thrombosis or hemorrhage.

Given the superior soft tissue contrast of magnetic resonance imaging (MRI), this modality can provide delineation of the tumor characteristics and its relationships with the affected nerve and surrounding structures more clearly. Typical hemangiomas exhibit uneven or short T1 signal and long T2 signal on MRI, and demonstrate a gradual filling pattern of contrast enhancement.

Two possible origins for haemangiomas are averred: reactivation of quiescent embrional haemgioblasts or neoangiogenesis that leads to the growth of new aberrant vessels [9]. An imbalance between angiogenic and inhibitory factors ultimately drives endothelial cell proliferation and increased vessel density, culminating in hemangioma formation [10]. Based on histological features, hemangiomas can be further classified including cavernous (blood-filled spaces lined by flat endothelium), arteriovenous (fetal capillary remnants with anomalous arteriovenous communication), and capillary (small vessels with thickened, muscularized walls) subtypes, with the latter being the most common [9].

Intraneural hemangiomas have been reported to involve digital nerve, ulnar nerve, median nerve, tibial nerve, superficial peroneal nerve, spinal nerve, etc, with the median nerve being the most commonly affected site. (**Table 1**) Most patients present with neurological symptoms related to the impacted nerve, such as numbness, pain, sensory deficits, and muscle atrophy. However, in the case described, the mediastinal intraneural hemangioma was an incidental finding, as the patient remained entirely asymptomatic prior to surgery. Upon 3 months of follow up, she complained of numbness in the right upper limb and tingling sensation during movement. It is speculated that the probable cause is the iatro-

genic nerve injury during the procedure. Case of increased numbness after operation of intraneural hemangioma has also been reported in the literature, the patient then received adjuvant radiotherapy, and such symptoms had disappeared by 2 years after radiotherapy [11].

There is no established consensus on the optimal management of intraneural hemangiomas, conservative treatment is usually less effective. Complete surgical resection although curative, carries a risk of iatrogenic nerve injury and surrounding structural damage. Subtotal resection may relieve symptoms with less collateral harm, carries a higher risk of recurrence [12]. In addition, a case report by Chatillion *et al.* demonstrated that symptoms could be relieved by radiation therapy, and mass shrinkage could be observed on imaging [11], suggesting it may be a viable alternative treatment option.

Table 1. Reviewing past cases of intraneural hemangiomas involving various nerves.

Ref.	Age	Sex	Involved nerve	Side	Symptoms	Size (mm)	Treatment	Post-treatment new deficits
Hadi [13]	64	F	extrafascicular left ulnar nerve	left	Pain paresthesia numbness	4 × 10	surgical excision	clawing persisted
Serdar [3]	12	M	Digital Nerve	right	numbness	15	surgical excision	no
Mohammed [14]	43	F	Median Nerve	left	numbness	9	surgical excision	no
Mastronardi [15]	41	M	Cauda equina	left	pain	15	surgical excision	no
Athanasios [16]	23	M	extradural lumbar nerve root	left	signs of radiculopathy	\	surgical excision	no
Xinyu Guo [17]	53	F	spinal nerve roots	right	muscular atrophy	25 × 20 × 20	surgical excision	no

4. Conclusion

Most patients with introneural hemangiomas have sensory or motor impairments in the affected nerves, MRI is the optimal modality to comprehensively evaluate these lesions, which typically appear as centripetal contrast enhancement. Surgical resection often yields a satisfactory outcome. However, the patient in this case found the lesion by chance, and the corresponding symptoms only developed after surgery. Notably, preoperative imaging did not include ultrasound or MRI, and no characteristic phleboliths were identified on CT, rendering the preoperative diagnosis more challenging compared to typical cases. This case highlights the importance of maintaining broad identification for uncommon mediastinal lesions. Careful postoperative monitoring is also crucial to

promptly identify and address any treatment-related sequelae.

Conflicts of Interest

The authors declare no conflicts of interest regarding the publication of this paper.

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