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Case Report / Olgu Sunumu

Primary pulmonary glomangioma

Primer pulmoner glomanjioma

Muhammet Reha Çelik,[1] Nurhan Şahin,[2] Ayşe Nur Akatlı,[2] Hakkı Ulutaş[1]

Departments of [1]Thoracic Surgery, [2]Medical Pathology, Medical Faculty of İnönü University, Malatya, Turkey

ABSTRACT

Glomangiomas are benign soft tissue tumors derived from the glomus bodies surrounding arteriovenous anastomoses. In this article, we report a 40-year-old female patient admitted to our clinic due to chest pain whose computed tomography of the chest revealed a solitary lesion of 4 cm in diameter. The lesion was removed with video-assisted thoracoscopic wedge resection. The histopathological diagnosis was reported as glomangioma. Glomus bodies almost never exist in the lung parenchyma; thus, pulmonary glomangiomas are extremely rare lesions. They can easily be confused with more common primary or metastatic lesions of the lungs. Risk of recurrence is low for this tumor and sublobar complete resection is the treatment of choice for definitive diagnosis and curative treatment.

Keywords: Glomangioma; glomus; pulmonary; video-assisted thoracoscopic surgery.

ÖZ

Glomanjiomalar, arteriyovenöz anastomozların etrafını saran glomus cisimlerinden kaynaklanan benign yumuşak doku tümörleridir. Bu yazıda, göğüs ağrısı nedeni ile kliniğimize başvuran ve bilgisayarlı göğüs tomografisinde 4 cm çapında soliter kitle saptanan 40 yaşında bir kadın hasta sunuldu. Kitle, video yardımlı torakoskopik kama rezeksiyon ile çıkarıldı. Histopatolojik tanı glomanjioma olarak bildirildi. Glomus cisimleri akciğer parenkiminde neredeyse hiç bulunmamaktadır; bu nedenle, pulmoner glomanjiomalar son derece nadir lezyonlardır. Daha sık görülen primer veya metastatik akciğer lezyonları ile kolaylıkla karıştırılabilir. Bu tümörde nüks riski düşük olup sublobar komplet rezeksiyon kesin tanı ve küratif tedavi için en doğru tercihtir.

Anahtar sözcükler: Glomanjioma; glomus; pulmoner; video yardımlı torakoskopik cerrahi.

Glomangiomas, also known as glomuvenous malformations, are considered to be variants of glomus tumor. Most of the cases are asymptomatic and generally occur more often during childhood. Unlike classic glomus tumors, which usually appear subungal, most predominate on the hand and forearm.[1] Glomus tumors are recorded to account for less than 2% of all soft tissue tumors. Since glomus bodies are rare or absent in the lung parenchyma, pulmonary glomangiomas are extremely rare lesions.[2] In this article, we report a case of primary glomangioma of the lung with chest pain.

CASE REPORT

A 40-year-old female patient admitted to our outpatient clinic with the complaint of chest pain. She was a non-smoker and her medical history was unremarkable. Laboratory data was within normal ranges. Imaging studies revealed a well circumscribed pulmonary lesion of 4.0 cm in diameter in her right lower lobe. A contrast-enhanced computed tomography (CT) of the chest demonstrated a solid lesion with regular margins and moderate density which was initially diagnosed as carcinoid tumor, radiologically (Figure 1). Preoperative fiberoptic bronchoscopy was performed and no endobronchial lesion was seen. Videothoracoscopic exploration showed an encapsulated mass, arising from right lower lobe and extending to the fissure through the intermediate bronchus. Thoracoscopic wedge resection of the adjacent lung within safe margins was performed with the removal of the mass. A written informed consent was obtained from the patient.

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Correspondence: Ayşe Nur Akatlı, MD. İnönü Üniversitesi Turgut Özal Tıp Merkezi, Tıbbi Patoloji Anabilim Dalı, 44280 Malatya, Turkey.
Tel: +90 422 - 341 06 60 e-mail: aysenurakatli@gmail.com

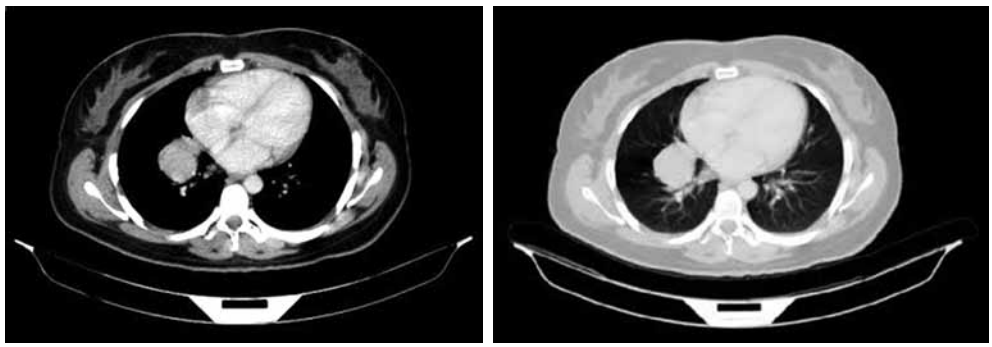

Figure 1. Right lower lobe solitary mass lesion is shown in preoperative chest computed tomography.

The resected specimen was placed in 10% formalin solution and sent to the pathology laboratory. The specimen was grossly 6x5 cm (Figure 2a), and cut surface was resembling a cavernous hemangioma. Microscopically, the lesion was composed of gaping veins with small, round nucleated uniform cells without mitotic activity in their walls and some solid areas of these uniform glomus cells (Figure 2b-2c). Immunohistochemically, glomus cells stained with vimentin, caldesmon, and smooth muscle actin (SMA) antibodies (Figure 2d). Type IV collagen demonstrated intracellular staining pattern (chicken-wire pattern).

Based upon the histomorphological and immunohistochemical findings, the case was reported as a glomangioma. The patient had an unremarkable postoperative course. Follow-up chest CT was negative for recurrent tumor and the patient remains free of disease 13 months after surgery.

DISCUSSION

Glomus tumors arise from the cells of arteriovenous anastomosis of the glomus bodies, or the Sucquet-Hoyer canal. Glomus cells have features of smooth muscle and are thought to be derived from the epithelioid

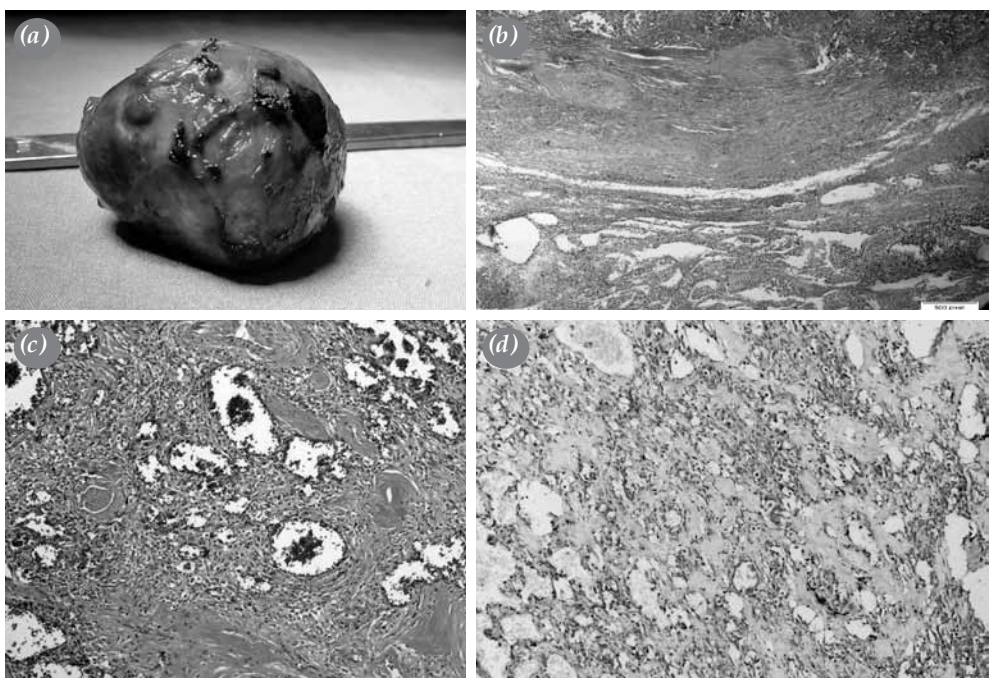

Figure 2. (a) Lesion was totally removed with adjacent lung tissue. Stapled lung tissue is apparent on left side. **(b)** Highly vascularized lesion adjacent to emphysematous lung tissue (H-E x 40). **(c)** Uniformly arranged tumor cells with clear cytoplasms and round nuclei (H-E x 100). **(d)** Smooth muscle actin (SMA) antibody immunoreactivity is shown in tumor cells (SMA x 100).

neuromuscular cells in the arteriovenous anastomosis of the glomus body and are associated with blood flow and temperature regulation due to their ability of contraction.[1,3-5]

Most glomus tumors occur as painful skin nodules in the upper extremities, subungual region of fingers and toes. Glomus tumors arising in the respiratory regions are extremely rare and the majority of the cases have been reported in the trachea.[3] Glomus tumors are well circumscribed lesions and composed of three components including glomus cells, blood vessels, and smooth muscle. Histopathologically, glomus tumors are subdivided into three categories based on the prominence of glomus cells (solid glomus tumor), vessels (glomangioma), and smooth muscle (glomangiomyoma).[3] The tumors present as either solitary lesions or as multiple lesions mimicking metastasis. Most of the glomus tumors are benign but rare cases of glomus tumors that demonstrate aggressive or malignant clinical and histological features have also been reported.[3,5-7] Folpe et al.[7] analyzed 52 unusual glomus tumors and suggested a classification as follows: malignant glomus tumor (glomangiosarcoma), glomus tumor of uncertain malignant potential, symplastic glomus tumor, and glomangiomatosis. They proposed that deep tumor location and size larger than 2 cm, or presence of atypical mitotic figures, or moderate to high grade nuclear atypia, and ≥5 mitotic figures/50 high-power fields should be considered as criteria for malignant glomus tumor. Tumors with high nuclear grade in the absence of any other malignant feature are considered as symplastic glomus tumor; tumors that lack criteria for malignant glomus tumor or symplastic glomus tumor but have high mitotic activity or large size or deep location only are defined as glomus tumor of uncertain malignant potential. They classified the tumors with excess glomus cells and histologic features of diffuse angiomatosis as glomangiomatosis.[7]

The median age of patients with primary pulmonary glomangioma was 43 to 45 years in the literature. Males are affected more than females (male to female ratio of 7:1).[8] Our case was a 40-year-old female patient. She was admitted to our hospital with chest pain. Although the pulmonary glomus tumors are generally larger than glomus tumors in soft tissue, ranging from 0.5 to 9.5 cm in diameter, the majority of the cases reported in the literature are asymptomatic. Pulmonary glomus tumors are usually detected incidentally on chest radiographs.[8]

Although glomangiomas are essentially benign lesions, the correct diagnosis has to be confirmed by the histopathological examination since glomangiomas do not present with specific findings in CT, magnetic resonance imaging (MRI) or 2-deoxy-2-[18F] fluoro-D-glucose positron emission tomography.[4] The characteristic morphological and immunohistochemical findings of glomangioma can be easily diagnosed by pathological examination.

The differential diagnosis for pulmonary glomus tumors should exclude carcinoid tumors, paragangliomas, hemangiopericytomas, smooth muscle neoplasms, cavernous hemangiomas, and metastatic glomus tumors.[4,8] It is difficult to distinguish a glomangioma from these tumors and metastatic nodules of the lung, because all of these appear under chest radiological examinations as round, well-delineated mass with strong enhancement.[9] However, these features of glomangioma on chest tomography scans usually distinguish it from lung cancer. Other benign lesions such as hamartomas or hydatid cysts may be differentiated by lesser degree of enhancement with CT or MRI scans.

Immunohistochemistry is useful in the differential diagnosis, as glomus cells express SMA, caldesmon and pericellular type IV collagen antibody. Glomangioma has negative immunoreactivity for desmin, CD34, cytokeratin, and S-100. Although carcinoid tumors have similar morphology with glomus tumors, they express immunohistochemical markers such as cytokeratin, chromogranin, synaptophysin, and CD56 which are negative in glomus tumors. Paragangliomas show immunohistochemical positivity with neuroendocrine markers and S-100, in contrast with glomus tumors. Glomus tumors can demonstrate a hemangiopericytomatous vascular pattern. But hemangiopericytomas are composed of spindle cells with elongated nuclei which are positive for CD34 and negative for SMA immunohistochemically and lack the uniform round cells that are seen in glomus tumors.[3] The tumor in our case was stained with SMA, caldesmon, and type IV collagen as in the literature.

Pulmonary glomus tumors in the lung are extremely rare. Most of the reported cases are different types of glomus tumors according to their morphological presentation and a few cases of primary glomangioma in the lung have been reported in the literature.[4]

Pulmonary glomangiomas have an excellent prognosis, and surgical excision is curative when a complete resection has been performed. Different surgical techniques have been employed for resection of such a benign tumor, including lobectomies due to

lack of diagnosis or size and location of the tumor.[3,9,10] Although a definitive diagnosis from frozen section is often not possible, with the exclusion of the several different malignant tumors, a pulmonary wedge resection should be adequate.

Due to uncertain behavior of such tumors and lack of specific characteristics in imaging studies, their frequency may be underestimated. It is important to report these cases, since surgical resection is necessary once they have been diagnosed and sublobar resections of the lung may adequately be performed to avoid larger surgical resections.

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