Solitary Arteriovenous Hemangioma of the Lung without Fistula: Report of a case and literature review

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Abstract:
Arterio-venous hemangiomas (also called vascular malformations) are uncommon lesions in general and very rarely reported as solitary lung tumors with no evidence of fistula formation or clinical evidence of shunting.

We report the case of a 49 year old Indian male who presented to our institution with a right hilar lung mass which was histologically proven to be a classical arteriovenous hemangioma (vascular malformation). The patient did not have clinical or radiological evidence of fistula or shunts formation.

Keywords: Arterio-venous hemangioma, Vascular malformation, Lung

Pathological Findings and Post-operative Follow-up:

The received specimen consisted of a right upper lobe of lung containing a well circumscribed, pale, firm and homogenous peribronchial tumor mass measuring 3.1 cm in its maximum diameter. The mass did not reach the pleural surface and the surrounding lung parenchyma was unremarkable apart from mild congestion. A few anthracotic peribronchial lymph nodes were also found.

Both frozen and paraffin-embedded histological sections showed a vascular tumor consisting of many benign medium-sized arterioles with a few scattered venules in between (Figure IA). At the periphery of the lesion several markedly dilated ‘feeder’ vessels were identified (Figures IA and IB). Some of these vessels were venous in origin and showed mild subintimal fibrosis. There were no thrombi in any of the described vascular channels and a histological diagnosis of arterio-venous hemangioma (vascular malformation) was made. Histological examination of the lymph nodes showed mild reactive changes only. The patient had an uneventful post-operative recovery and he remains well after three months of follow-up.

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Figure 1A and 1B: Solitary arterio-venous hemangioma of lung: CT of the thorax after intravenous injection of contrast agent coronal and axial: Well defined mass anterosuperior to the right upper lobe bronchus 3 cm in its maximal dimension highly enhanced post intravenous injection not associated with bronchial narrowing, bronchial masses or distal pulmonary collapse consolidation.
Case Report:

A 49 year old Indian male presented to our Chest Clinic with a history of persistent cough which did not respond to the usual medications. There was no significant past medical history and his physical examination, routine biochemistry and hematology profiles were unremarkable, and no clinical or radiological evidence of vascular shunts could be detected.

Radiological investigations, which included computed tomography examination of the chest, revealed the presence of a relatively well-circumscribed right hilar lung mass which was impinging on the right main and upper lobe bronchus, and measured 3 centimeters in its maximum diameter.

Because of the clinical and radiological suspicion of this lesion being a benign neoplasm in addition to negative sputum cytology and bronchoscopy results, a decision to refer this patient for surgery was taken, and a subsequent resection of the right upper lobe of lung with sampling of adjacent peribronchial and mediastinal lymph nodes was done, and sent for histological assessment.

Discussion:

Arteriovenous hemangiomas are uncommon lesions which usually occur more frequently in the limbs, head and neck areas, of adolescents and young adults\(^1,3\).

The deep seated lesion may represent congenital malformation, and can be associated with severe clinical symptoms, which include heart failure or Kasabach-Merrit Syndrome\(^3\). The superficial types of these lesions are also known as cirrhotic aneurysms or acral arteriovenous tumors\(^2\). They are typically seen in the skin of the head and neck (especially the lip) of middle-aged adults, and present as a small red-blue papules.

Shunting is usually not a major feature, and symptoms include mild pain and occasional intermittent bleeding. Superficial cutaneous changes associated with deep arteriovenous haemangiomas can, however, mimic Kaposi's sarcoma clinically and histologically, and have been named pseudo-Kaposi's sarcoma or acralangiodermatitis\(^4\).

Histologically the main characteristic of this tumour is a proliferation of uniform small to medium-sized veins and arterioles, sometimes with inconspicuous lumina. The intervening stroma is usually myxoid. Endothelial cells may be plumper than normal but not atypical. They stain for factor eight related antigen, CD34 and Ulex Europa\(^1,2,5\). In addition, these vessels have a fibro-muscular wall which contains elastic fibres but with no definite elastic lumina\(^5\). In most reported cases the vessels have the characteristics of veins; however, our case showed a predominance of small arterioles mainly in the central parts of the lesion.

To the best of our knowledge this case represents a very rare example of a classical and solitary arteriovenous haemangioma of the lung. The lack of arteriovenous fistula, absence of shunting and the predominance of the arteriolar proliferation in our patient's lesion make this case almost unique as most cases which are reported in the literature\(^7\) are associated with fistula formation\(^6\), emboli\(^6\), aneurysms\(^7\), Osler-Weber-Rendu disease\(^8\), hepatic fistulas\(^8\) or are labeled as bronchial arterioma\(^9,10\).

Finally, the issue of whether these lesions should be classified as a neoplasm (arteriovenous hemangioma) or a vascular malformation is still controversial and in need of further studies probably at the molecular genetics level.
Conclusion:

In patients presenting with well defined and single hilar or bronchial lung lesions, the rare possibility of arteriovenous hemangioma should be considered, investigated and/or at least included in the differential diagnosis, even in the absence of clinical and radiological evidence of vascular shunt formation.

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