CASE REPORT

A rare case of congenital complex pulmonary AV fistula

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SUMMARY

A 15-year-old boy presented with central cyanosis with clubbing and dyspnoea on exertion. Cardiovascular examination did not reveal any abnormality. ECG was normal. Chest X-ray showed a normal sized heart with rounded opacities of variable size in the left upper lung field. Two-dimensional echocardiographic examination was normal. CT angiography showed a large complex lesion composed of serpiginous tubular structures involving the left upper and lingular lobe, suggestive of racemose tangle of blood vessels. A diagnosis of large complex arteriovenous (AV) fistula involving the left upper and lingular lobe was performed. This case reports a rare case of complex pulmonary AV fistula.

BACKGROUND

This case holds clinical importance as complex pulmonary arteriovenous (AV) fistula is a rare entity. The rarity of this disease and the severity of symptoms raises concerns as to the importance of this entity.

CASE PRESENTATION

A 15-year-old boy presented with cyanosis and dyspnoea on exertion. There was no history of haemoptysis, chest pain, headache, liver disease or infection. On examination, he had central cyanosis with clubbing (figures 1 and 2). Peripheral pulses were not bounding. There was no evidence of any cutaneous telangiectasia. Cardiovascular examination did not reveal any right to left shunt, cardiomegaly or any other abnormality. ECG was normal (figure 3). Chest X-ray showed a normal sized heart with rounded opacities of variable size in the left upper lung field (figure 4). Two-dimensional echocardiographic examination was normal. CT angiography showed a large complex lesion composed of serpiginous tubular structures involving the left upper and lingular lobe suggestive of racemose tangle of blood vessels (figure 5). A cord-like band was seen extending from the lesion to the hilum (figure 6). Left pulmonary artery, left upper lobe artery and branches of the lingular lobe were dilated (figure 7). A diagnosis of large complex AV

fistula involving the left upper and lingular lobe was performed.

Since the patient refused any intervention, he is being followed up conservatively.

DISCUSSION

Pulmonary AV fistula is a very rare anomaly. Most patients with pulmonary arteriovenous malformations have hereditary haemorrhagic telangiectasia (HHT),¹ an autosomal dominant disease; or have acquired pulmonary AV fistula due to liver disease or infections such as tuberculosis.^{2 3} In this case all of these possibilities were particularly excluded. Also, most pulmonary arteriovenous malformations are found in the lower lobes. This case reports a rare case of congenital pulmonary AV fistula not associated with HHT and involving the left upper and lingular lobe.

Figure 1 Showing cyanosis and clubbing of fingers.

Figure 2 Showing cyanosis and clubbing of toes.

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Figure 3 Normal ECG. **Figure 4** Chest X-ray showing normal sized heart with rounded opacities of variable size in the left upper lung field. **Figure 6** CT angiography images showing a cord-like band extending from the lesion to the hilum. **Figure 5** CT angiography images showing a large complex lesion composed of serpiginous tubular structures involving the left upper and lingular lobe suggestive of racemose tangle of blood vessels. **Figure 7** CT angiography images showing dilation of the left pulmonary artery, left upper lobe artery and branches of the lingular lobe.

Learning points

- ▶ Congenital pulmonary arteriovenous fistula is a very rare finding.
- ▶ It can occur without any other associated condition.
- ▶ It can also less commonly involve the upper lobe of the lung.

Competing interests None.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

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