CASE REPORT

Endovascular treatment of isolated systemic arterial supply to normal lung with coil and glue embolisation

^{1,2}G ANIL, MD, FRCR, ^{1,3}M TANEJA, MD, FRCR and ¹A-G-S TAN, MBBS, FRCR

¹Department of Radiology, Changi General Hospital, Singapore, ²Department of Diagnostic Imaging, National University Hospital, Singapore, and ³Interventional Radiology Centre, Department of Radiology Singapore General Hospital, Singapore

ABSTRACT. Surgery is the standard treatment for the extremely rare pathology of isolated anomalous systemic arterial supply to normal lung (ISSNL). We describe our experience with this anomaly in a 29-year-old male presenting with recurrent haemoptysis that was successfully treated with a combination of metallic coils and cyanoacrylate glue. In addition to contributing to the extremely limited data on endovascular therapeutic options in ISSNL, we also intend to raise the awareness among endovascular therapists of the need to be cautious of and preserve the radiculomedullary/pial branches arising from an anomalous artery before embolising it.

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Anomalous systemic arterial supply to normal lung, with no associated bronchopulmonary mass/cyst or abnormality in the bronchial tree is the rarest of the pathologies in the “sequestration spectrum” [1]. It is usually treated surgically [2]. We present an adult case of a large anomalous systemic arterial supply to basal segments of the left lower lobe embolised with N-butyl cyano-acrylate glue (NBCA) and metallic coils.

Case report

A 23-year-old male presented with complaints of recurrent haemoptysis in the previous week. He mentioned having similar episodes on several occasions since childhood which had been treated as bronchiectasis. There was no history of significant perinatal illness.

A chest radiograph showed retrocardiac tubular densities. A post-contrast CT of the thorax revealed a large artery (12 mm in diameter) arising from the lower descending thoracic aorta that supplied the posterobasal and laterobasal segment of the left lower lobe with no supply from the left inferior pulmonary artery (Figure 1). The anomalous artery (AA) had a swan-neck like configuration; after its first downward turn, it looped cranially and as it curved down a second time it ramified into multiple large capacious branches corresponding to the retrocardiac shadows seen on the radiographs. There was no abnormal communication between the AA and the pulmonary artery/vein. The bronchopulmonary segments involved were plethoric (Figure 1d). There was normal drainage through a dilated inferior pulmonary vein. There were no inflammatory changes. The tracheobronchial tree and the rest of the lungs were unremarkable. The CT

scan permitted the diagnosis of isolated systemic arterial supply to normal lung (ISSNL). A bronchoscopy confirmed the bronchial anatomy was normal.

The therapeutic options were discussed with the patient, ranging from continued monitoring to surgery and transarterial embolisation (TAE) and the possible associated complications. The patient chose the endovascular option with full awareness of the lack of adequate data on the long term outcome of TAE.

An angiogram confirmed the CT findings (Figure 2a,b). Through a 4 French catheter (Simmons Type 1; Cordis Miami, FL) hooked into the AA, a 2.7 French microcatheter (Progreat; Terumo Tokyo, Japan) was used coaxially to cannulate its branches. Every first order branch was embolised back with metallic coils [0.018 inch; 22 coils; Nester coil, (William Cook Europe, Oslo, Norway) 3–8 mm diameter and Complex helical, (Boston Scientific, Cork, Ireland) 5–6 mm diameter and 2 ml of NBCA glue [Histacryl; B Braun (Aesculap), Melsungen, Germany] diluted in 2 ml of lipiodol. The glue was injected into a loose scaffold of metallic coils with a compact block of coils proximally and distally. A long proximal stump of the AA was left unembolised because radiculomedullary and radiculopial branches arose from here. Completion angiograms showed total cessation of aberrant pulmonary arterial flow (Figure 2c). The patient had chest discomfort for 2 days following the procedure and could be discharged on the third day.

At 3 month and 6 month follow-up, the patient was asymptomatic with no pulmonary infarction on the chest radiographs (Figure 2d). Thereafter, he was lost to follow-up.

Discussion

According to the Pryce nomenclature from 1946, such a case of ISSNL would be categorised as Type 1

Address correspondence to: Dr Gopinathan Anil, Department of Diagnostic Imaging, National University Hospital, 5 Lower Kent Ridge Road, Singapore 119074. E-mail: ivyanil10@gmail.com

Figure 1. In these contrast enhanced CT images the anomalous artery (white arrow) arising from the thoracic aorta and supplying the left basal segments is demonstrated in three orthogonal planes (a–c). (d) Note the ill-defined, faint airspace haziness and increased vascularity in the affected basal segment of the left lower lobe.

sequestration [3]. However, unlike the original definition of sequestration, this patient had no associated bronchopulmonary mass, cyst or abnormality in the development of bronchial tree [1, 3]. To adequately classify such cases, the concept of “sequestration spectrum” was introduced. At one end of the spectrum normal vessels supply abnormal lung and abnormal vessels supply normal lung at the other [1]. Alternatively, the term “pulmonary malinosculature” is used for the entire range of congenital abnormal connections or openings of one or more of the four components of lung tissue, *i.e.* airway, arterial supply, venous drainage and parenchyma. The abnormalities of the airway, arterial

supply, or both are classified as tracheobronchopulmonary malinosculature, arterial malinosculature and bronchoarterial malinosculature, respectively. The classification is completed by describing the associated findings of venous drainage and lung parenchyma [4].

The usual pattern in ISSNIL is an AA arising from the descending aorta and supplying the left lower lobe. Rarer patterns include AA arising from other arteries such as the coeliac trunk, involvement of other regions of lung such as the upper lobe or the right lung and simultaneous systemic and pulmonary arterial flow into the same segments [2]. Although the aetiology of systemic arterialisation

Figure 2. (a) The thoracic aortogram shows the origin and tortuous course of the anomalous artery. (b) Selective injection into the anomalous artery demonstrates the branching pattern and parenchymal blush of the subtending segments of lung. (c) Selective injection into the trunk of the anomalous artery post-embolisation with coils and NBCA glue shows no parenchymal enhancement but the spinal artery is preserved. (d) Follow up radiograph at 3 months does not show any evidence of pulmonary infarction.

is unknown, the most likely explanation is the persistence of intersegmental arteries that connect the embryonic dorsal aorta with pulmonary parenchyma [5].

ISSNL is often asymptomatic and may get detected incidentally on radiographs or present as continuous murmurs mimicking patent ductus arteriosus. The pulmonary arterial branches supplied by the AA cannot

withstand the high systemic arterial pressures and they tend to bleed, presenting as haemoptysis, although they are not usually massive [2]. Exertional dyspnoea and cardiac failure from large left to left shunt is uncommon.

In the diagnosis and characterisation of ISSNL multi-slice CT has largely replaced catheter angiography [6]. A CT scan can differentiate ISSNL from other causes of

systemic arterialisation of the lung such as pseudosequestration, where chronic inflammation initiates large anastomoses between the pulmonary and systemic arteries. Systemic supply from multiple arteries in the vicinity and scarring reveals the diagnosis in such cases. Severe attenuation of the pulmonary arteries in Takayasu's arteritis with the consequent parasitisation of systemic arteries by the subtending bronchopulmonary segments may mimic congenital systemic arterialisation. In such cases the bronchial or phrenic arteries are usually hypertrophic. The systemic arterial supply seen with pulmonary arteriovenous malformations is either secondary to a shunt induced ischaemia or in response to surgery, but identified by their abnormal venous drainage [7].

Lobectomy/segmentectomy with dissection of the AA is the standard treatment [8]. Other surgical options include simple ligation of the systemic artery [9] or switching of the AA from the aorta to the pulmonary trunk (in children). In the last decade, there have been some reports on TAE as a therapeutic option for ISSNL [8]. According to a recent review, only six patients have undergone TAE for ISSNL [10]. All these cases achieved therapeutic success without major complications. Although TAE entails a theoretic risk of pulmonary infarction and secondary infection/fistulae, there are no documented complications that required additional management.

The use of glue to stabilise coils and reduce the number required to achieve adequate embolisation, is an established practice among interventionalists with experience in handling liquid embolising agents. In this case, we extrapolated this advanced technique to good effect. To the best of our knowledge, this is the first instance of TAE performed for ISSNL using both glue and metallic coils. The published English language literature only describes the use of metallic coils for this purpose. In our patient, the inherent vascular anatomy with upward course of the main feeding artery, its termination into multiple large branches and the spinal artery arising a few centimetres proximal to its ramification posed unique challenges; namely, (1) precluding the use of Amplatzer Vascular Plug (AGA Medical Corporation, Plymouth, MN) to occlude the main artery, (2) occluding the stump without dealing with the first order arteries ran the risk of proximal coil migration (hence we had to selectively occlude each of the large first order branches). Compact distal coiling was critical to prevent the glue from freely percolating into the distal arterial bed that could increase the risk of pulmonary infarction. Similarly, controlled injection of the glue was essential to prevent retrograde flow into the descending aorta and its obvious disastrous complications. The fortuitous presence of a long segment of AA before it branched also provided us with a reasonable margin of safety against such a mishap. The inadvertent embolisation of radiculomedullary/pial branches was avoided by ensuring that when the glue was injected, the microcatheter tip is well beyond the origins of these branches. The lipiodol in the glue mixture allows

good visualisation and helps in facilitating a controlled injection as well as determining the end point of embolisation. The use of NBCA in such vital structures as the thoracic aorta and lungs with the precarious anatomy of the vessel to be embolised warrants the operator to have adequate experience and skill in handling this unforgiving embolising agent.

Conclusion

In addition to endorsing the utility of multislice CT in accurately diagnosing and characterising ISSNL, this report adds to the limited data available on the endovascular therapeutic options available for this pathology. In experienced hands, the technique of NBCA trapping in a loose scaffold of metallic coils between a compact stack of coils may be a useful improvisation even in complex clinical situations, as demonstrated in the featured case.

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