CASE REPORT

Spontaneous abdominal arteriovenous fistula and polysplenia in a child presenting with brain abscess

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SUMMARY

Spontaneous arteriovenous communications below the diaphragm is a very rare condition. Its association with polysplenia has perhaps not yet been reported in children. We reported a case in a 9-year-old boy presenting with acute onset of fever, vomiting, headache, seizures and altered sensorium. A CT scan revealed a large occipital abscess which was drained surgically. On examination he had marked central cyanosis and grade III clubbing of fingers and toes. Further investigations led to the discovery of a very rare finding of abdominal arteriovenous malformation associated with multiple-enhancing structures around the splenic bed suggestive of polysplenia. We believe that the most likely cause of the brain abscess was paradoxical embolism through the abdominal arteriovenous communication. Unfortunately the parents did not consent to a surgical correction of the malformation, realising the risks involved in the procedure. Subsequently the boy was lost in follow-up.

BACKGROUND

Abnormal arteriovenous communications may be congenital or acquired. The acquired variety is usually traumatic in origin. In rare cases it may be spontaneous due to disease of an arterial wall. This may result in the formation of arterial aneurysm, which in turn may perforate into the contiguous veins.

James Syme in 1831¹ first reported a case of ruptured aortic aneurysm producing aortocaval fistula. Following this very few cases of spontaneous arteriovenous communications were reported. Most of these were located in the thorax. Hence, a spontaneous and abnormal communication between an artery and a vein below the diaphragm is an exceptionally rare entity.

We reported this unique case of a 9-year-old boy. He presented to us with sudden onset of fever, vomiting, seizure and altered sensorium. The fever followed mild coryza. It was high grade and was associated with rigor. There was severe headache and recurrent vomiting along with fever spikes. The child developed slurring of speech and altered sensorium. Two days prior to the hospital admission he had two episodes of generalised seizures. The child was barely responding to verbal commands and was unable to walk or sit alone. The child did not have any obvious congenital deformities or dysmorphic features. There was no significant history apart from minor respiratory tract infections.

An urgent CT scan of the brain revealed a large solitary abscess located in the occipital region of his

brain. The abscess was drained by neurosurgical intervention. However, clinical examination also revealed that the child had marked central cyanosis and clubbing of fingers and toes. On further questioning parents stated that they noted these features since his early childhood. The parents consulted the local doctor. He had some investigations conducted at that time. Those investigations included a pulmonary angiogram and an echocardiogram with colour Doppler studies. They were reported as normal. We arranged for further investigations to delineate the underlying cause. A dynamic (triphasic) CT scan of the abdomen revealed abnormal arteriovenous communication along with multiple-enhancing structures in the splenic bed (figure 1). Thus, the very rare diagnosis of abdominal arteriovenous fistula with polysplenia was found. The polysplenia in this case was a purely incidental finding. This was unlikely to produce any of his symptoms. In many cases the polysplenia do not cause significant clinical symptoms. The diagnosis is only confirmed by suggestive features in specialised dynamic vascular imaging techniques. The abnormal arteriovenous communication in all likelihood was spontaneous in origin. This is more so in the absence of any significant history or trauma. However, in some cases, these may be confused with the congenital variety of arteriovenous fistula. In any case this was an exceptionally rare association, probably not reported in children till now. Moreover, in the absence of any other predisposing factors, we strongly believe that the brain abscess was caused by paradoxical septic embolisation through this abnormal communication. The above reasons make this case worth reporting.

INVESTIGATIONS

On admission: haemoglobin 17.6 g%, total white cell count 21 200, neutrophil 71%, C reactive protein 11.2 (normal <1), malaria parasite: absent.

Liver function test: Normal.

CT scan of brain: large solitary abscess in the occipital region. No significant midline shift (figure 2).

Mantoux test: Negative. *Cerebrospinal fluid (CSF)*

STUDY: cell <5, protein and sugar within normal range.

Electrolytes, urea and creatine: Normal, blood cultures: negative, *thyroid profile:* normal.

Chest X-ray: normal, immunoglobulin assay: normal, *arterial blood gas:* PaO₂<50 mm Hg despite hyperoxia, *methaemoglobin level:* normal.

Aspirated material from drained abscess grew *Staphylococcus aureus* in large number.

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Figure 1 Abnormal arterio venous communication.

Figure 2 Occipital brain abscess.

Figure 3 Abnormal arterio venous communication with multiple enhancing structures in splenic bed.

Echocardiogram with colour Doppler: no structural anomaly, good biventricular function.

Pulmonary angiogram was normal.

CT Scan of abdomen (dynamic 3-phase study): Liver, biliary radicals and porta hepatis were normal. Gallbladder long and folded, common bile duct normal, pancreas and main pancreatic ducts were normal; kidneys and pelvic calyceal systems were normal on both sides.

Multiple-enhancing structures in the splenic bed suggestive of polysplenia (figure 3).

Vascular study: aorta normal; upper part of inferior vena cava was opacified at the arterial phase suggesting aortocaval communication.

Splenic and renal veins were prominent; portal vein was not visualised, hepatic veins normal.

A sac-like structure was noted at the level of renal vein lateral to the aorta on the right side and compressing the inferior vena cava. This sac-like structure receives supply from distended abnormal vessels just cranial to the left renal vein and then drains in to the inferior vena cava.

Blood reports on day 5 following surgical evacuation of the brain abscess; haemoglobin 15.6 g%, total white cell count 7800, neutrophil 53%, C reactive protein 1.4.

DIFFERENTIAL DIAGNOSIS

At the time of presentation the child had symptoms suggestive of infection of the central nervous system. These included fever,

vomiting, seizures and altered sensorium. Hence, the possibility of meningoencephalitis was considered. A CT scan and lumbar puncture were performed. The CT scan of the brain clearly revealed a large solitary abscess in the occipital region of the brain. The CSF study was normal.

On clinical examination, the child had central cyanosis and grade 3 clubbing of his fingers and toes. So we considered the differential diagnoses for central cyanosis and clubbing.

Most pulmonary causes (ie, pneumonia, pulmonary oedema, obstructive airway disease) were excluded by a normal chest X-ray and ear nose and throat examination. Structural cardiac lesions and anatomical shunts were excluded by a normal echocardiogram and colour Doppler studies. Pulmonary arteriovenous fistula and intrapulmonary shunts were excluded by normal pulmonary angiogram. The possibility of haemoglobin abnormalities including methaemoglobinaemia that may cause cyanosis were excluded by appropriate investigations.

After exclusion of these possibilities we investigated his abdomen. The ultrasound revealed anomalies including the possibility of polysplenia and vascular anomalies. Hence, a dynamic, three-phase abdominal CT scan was performed. This revealed the sac-like abnormal structure. The visualisation of the inferior vena cava at the arterial phase clearly suggested arteriovenous communication in the abdomen.

This abnormal arteriovenous communication was spontaneous in origin as there was no significant history to explain its aetiology. However, as the parents noted the bluish

discolouration in the child since early childhood, we considered the congenital cause as well.

Therefore, we examined the child meticulously but could not find any other evidence of congenital deformity. The child also had numerous advanced investigations carried out to delineate the underlying cause of his clinical symptoms. These involved CT of the brain and abdomen, echo with colour Doppler studies as well as pulmonary angiogram. Apart from the incidental findings strongly suggestive of polysplenia, nothing else was diagnosed.

Hence, we suggested the diagnosis of spontaneous abdominal arteriovenous fistula with polysplenia in this case.

TREATMENT

Investigations performed at admission clearly showed the large occipital space occupying lesion. The child also had central cyanosis and his oxygen saturation was between 80% and 90%. Hence very high risk consent was obtained from the family. The occipital lesion was drained by appropriate neurosurgical drainage. The pus was very thick and it grew *S aureus*. Initially the child was treated with intravenous ceftriaxone and vancomycin injection. Subsequently ceftriaxone was replaced by injection meropenam. The intravenous broad spectrum-antibiotic regime was continued for 2 weeks following the surgery. For the initial 48 h following surgical evacuation of the brain abscess, inotropic support was needed to maintain normal blood pressure. Other supportive treatment including administration of fresh frozen plasma and packed red blood cell was given immediately following the operation.

OUTCOME AND FOLLOW-UP

The child regained normal sensorium following successful drainage of the brain abscess. His symptoms subsided and clinical and laboratory signs of infection disappeared. Subsequent investigations revealed the presence of abnormal arteriovenous connection inside his abdomen. We explained to the family that this was the cause of his underlying problem. We also suggested further investigations including selective angiogram of the affected vessels before surgical correction at a specialised centre. However, the high mortality and poor prognosis associated with this procedure was also explained to them. We referred the child to a higher and the best equipped centre to perform the vascular surgery. There also the family was counselled in detail about the risks involved in undertaking the appropriate surgical intervention. Considering the risks involved in such a procedure, the parents decided not to undertake any further investigation and subsequent surgery. They realised the implication of their decision. Later on the child was lost in follow-up.

DISCUSSION

Since its original description by Syme¹ in 1831, aortocaval communications have rarely been reported. Until 1930, 75 instances of spontaneous communications between aorta and superior vena cava²⁻⁶ were noted. Spontaneous communications below the diaphragm are exceptionally rare. Matas⁷ in 1909 stated that of 114 such cases, only 20 involved abdominal aorta and inferior vena cava. Reid⁸ in 1925 could not add any further references. Therefore, aortocaval fistula is a very rare complication of spontaneous abdominal aneurysm rupture. The incidence is only 2-4%. However, the mortality rate of this condition is approximately 30%.⁹ Surgical repair of aortocaval fistula was first tried in 1935.¹⁰ Unfortunately the patient died soon after the surgery. Twenty years later Cooley¹¹ reported the first successful repair in 1955. In 1999 Tamiyasu and Tokunaga¹² reported successful repair of multiple aortocaval fistula in the abdomen of a patient

with Ehler Danlos syndrome. It is very important to note that none of these cases mentioned were from paediatric age group.

The abnormal communication between the artery and vein anywhere in the body may give rise to clinical signs such as central cyanosis and clubbing. The exact diagnosis, however, may remain unnoticed for years. This may be applicable for congenital cases as well. A high degree of clinical suspicion and specialised investigations are required to pinpoint the exact diagnosis. Contemporary literature searches reveal very few such reports of congenital aortocaval fistula involving the superior vena cava.¹³⁻¹⁴ Kuint *et al*¹⁵ in 1998 reported a congenital aortocaval fistula in the newborn period.¹⁵

In our exhaustive literature search we did not find a single report of abdominal arteriovenous communication associated with radiological findings of polysplenia in a child. The diagnosis may not even be obvious in normal CT scan or MRI. Only a dynamic triple phase study may suggest the abnormal filling or opacification of the veins in the arterial phase. This strongly suggests abnormal arteriovenous communication. Obviously this needs to be confirmed by selective angiogram of the involved vessels before actual surgical correction.

This abnormal connection may give rise to the right to left shunt. Thereby, a septic focus in the lower extremities can generate septic embolism. This may pass through the abnormal communication and bypass the pulmonary circulation. Hence, this form of embolisation may cause abscess in target organs such as brain. Laporte *et al*¹⁶ in 2010, described aorto caval fistula as an uncommon cause of paradoxical embolism. However, in our case we firmly believe this as the most likely cause of the brain abscess in this unfortunate child.

Learning points

- ▶ Abnormal arteriovenous communications below the diaphragm are exceptionally rare particularly in children.
- ▶ Despite clinical signs such as cyanosis and clubbing they may remain undiagnosed for years.
- ▶ Because of the presence of right to left shunt, they may be associated with paradoxical embolisation.
- ▶ They need to be diagnosed on the basis of high index of clinical suspicion and specialised investigations.
- ▶ The surgical corrections of these are associated with very poor prognosis and high mortality.

Contributors Both the authors were involved in the management of the case and actively contributed in the preparation of the manuscript.

Competing interests None.

Patient consent Obtained.

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REFERENCES

- 1 Syme J. Case of spontaneous varicose aneurysm. *Edinb Med J* 1831;36:104-5.
- 2 Pepper W, Griffith JPC. Varicose aneurysm of the aorta and superior vena cava. *Am J Med Sci* 1890;100:329.
- 3 Humphry L. Aneurysm of the aorta communicating with the superior vena cava. *Br Med J* 1910;2:1046.
- 4 Shennan J. Spontaneous arterio venous aneurysm in thorax. *Edinb Med J* 1925;32:325.
- 5 House SJ, Goodpasture EW. Spontaneous arterio venous aneurysm in the thorax. *Am Heart J* 1927-1928;3:682.
- 6 Packard M, Weshler HF. Arterio venous aneurysm between aorta and superior vena cava, Case report. *Am Heart J* 1930-1931;6:281.
- 7 Matas R. Keen's surgery, V, 290 ff. 1909.
- 8 Reid MR. Abnormal arterio venous communication, acquired & congenital. *Arch Surg* 1925;10:996.

- 9 Alexander JJ, Imbembo AL. Aorta vena caval fistula. *Surgery* 1989;105: 1–12.
- 10 Lehman E. Spontaneous arterio venous fistula between the abdominal aorta and inferior vena cava: case report. *Ann Surg* 1938;108:697–700.
- 11 Cooley D. Discussion of Javid H, Dye WS, Grave JW, Julian OC; Resection of ruptured aneurysm of the abdominal aorta. *Ann Surg* 1955;142:623.
- 12 Tamiyasu N, Tokunaga H. Multiple aorto caval fistula associated with a ruptured abdominal aneurysm in a patient with Ehlers- Danlos syndrome. *Jpn Circ J* 1999;63:564–6.
- 13 Kiatchoosakun S, Wongvipaporn C, Silarak S, *et al.* Congenital aorto caval fistula to the superior vena cava, a case report. *J Med Assoc Thai* 2003;86:381–4.
- 14 Oomman A, Mao R, Krishnan P, *et al.* Congenital aorto caval fistula to the superior vena cava. *Ann Thorac Surg* 2001;72:911–13.
- 15 Kuint J, Bilik R, Heyman Z, *et al.* Congenital aorto caval fistula in the newborn: a case report. *J Paediatr Surg* 1998;33:743–4.
- 16 Laporte F, Olivier A, Groben L, *et al.* Aorto caval fistula: an uncommon cause of paradoxical embolism. *J Cardiovasc Med (Hagerstown)* 2010. Source: Department of Cardiology, University Hospital, Van doeuve-les-Nancy, France.

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