Splenic abscess and perisplenic hematoma complicating Osler–Weber–Rendu disease

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ABSTRACT

A 52-year-old diabetic gentleman was referred to our center with a 3-week history of fever, left-sided abdominal pain, and progressive breathlessness. He also had history of recurrent epistaxis since childhood. Contrast-enhanced computerized tomography chest and abdomen revealed a splenic abscess, left pulmonary arteriovenous malformation, and left pleural effusion. He was managed conservatively with intravenous antibiotics and an antifungal. A repeat imaging was done after 3 weeks which showed resolution of abscess but an increase in the size of the perisplenic hematoma. An ultrasound guided pigtail catheter was inserted into the peri-splenic hematoma and it was drained. He had also developed an acute cerebellar infarct detected on magnetic resonance imaging of brain, which also showed other chronic infarcts of varying age. A diagnostic nasal endoscopy revealed multiple telangiectasias, and Osler–Weber–Rendu disease (hereditary hemorrhagic telangiectasia [HHT]) was diagnosed according to Curacao criteria. Symptomatic splenic involvement may be a rare manifestation of HHT.

Key words: *Cerebrovascular accident (CVA), Hereditary hemorrhagic telangiectasia, Pulmonary arteriovenous malformation, Spleen, Stroke*

Hereditary hemorrhagic telangiectasia (HHT; Osler–Weber–Rendu syndrome) is an autosomal dominantly inherited disorder with a prevalence of 1 in 10,000 to 1 in 5000 [1]. However, most of the patients are unaware of their diagnosis of HHT [2]. The diagnosis of HHT is made clinically on the basis of the Curaçao criteria, established in June 1999 by the Scientific Advisory Board of the HHT Foundation International, Inc. [3]. The four clinical diagnostic criteria are as follows: (1) Epistaxis, (2) telangiectasias, (3) visceral lesions arteriovenous malformations (AVMs), and (4) family history (a first-degree relative with HHT). Our patient had a splenic abscess and a perisplenic hematoma. After an extensive search of the literature, splenic involvement in HHT was found to be very rare.

CASE REPORT

A 52-year-old gentleman was referred to our center with fever, left-sided abdominal pain, and progressive breathlessness for the past 3 weeks. He also had a background of Type 2 diabetes mellitus, controlled on oral antidiabetic drugs, recurrent epistaxis since childhood and strong family history of epistaxis. Clinical examination revealed tachycardia, tachypnea, and signs suggestive of a left-sided pleural effusion with left-sided abdominal tenderness.

Our laboratory investigations revealed leukocytosis, microcytic hypochromic anemia, thrombocytosis, elevated inflammatory markers, a C-reactive protein of 289 mg/dl (<3 mg/dl), an ESR of 110 mm/h (0–22 mm/h), and procalcitonin of 7.8 ng/ml (0.15–2 ng ml). Liver and renal functions were within normal limits. Serology for hepatitis B and C, HIV 1 and 2, Brucella, and Leptospira was negative. WIDAL was negative for typhoid. Blood and urine cultures were of no growth. Tuberculosis was ruled out by cartridge-based nucleic acid amplification test. A contrast-enhanced computerized tomography of the chest and abdomen showed a small 2.9 cm × 1.3 cm multi-septated abscess involving the lower pole of the spleen with a large peri-splenic hematoma, left upper lobe pulmonary AVM, and bilateral pleural effusion (Figs. 1 and 2).

He was put on intravenous antibiotics and an antifungal. Bacterial culture from splenic hematoma was no growth. Two-dimensional ECHO was normal, while contrast ECHO was positive and suggestive of pulmonary AVM. A pleural fluid study was suggestive of exudative effusion, and tuberculosis was ruled out. A surgical opinion was taken and he was planned for splenectomy if his clinical condition worsened and he was continued on conservative management.

2 weeks later, he started to develop dyspnea again and so a repeat CT abdomen was done which showed complete resolution of the abscess but an increase in the size of the perisplenic hematoma as compared to previous CT, with a rent in left hemidiaphragm (Fig. 3). A ultrasound-guided Pigtail was inserted into the perisplenic hematoma. He later developed dysarthria, dysphagia, and nasal regurgitation, for which magnetic resonance

imaging + MR angiogram of the brain was done, which showed acute infarct of the cerebellum (Fig. 4) with old chronic cerebral, pontine, and cerebellar infarcts.

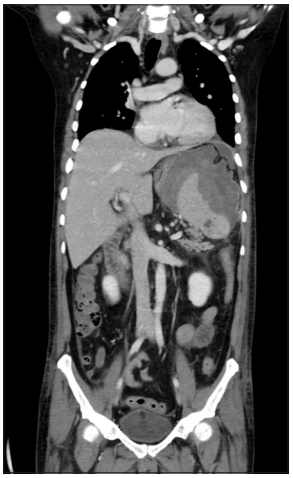

Figure 1: Contrast-enhanced computerized tomography showing a 2.9 cm × 1.3 cm multi-septated abscess involving the lower pole of the spleen with a large perisplenic hematoma

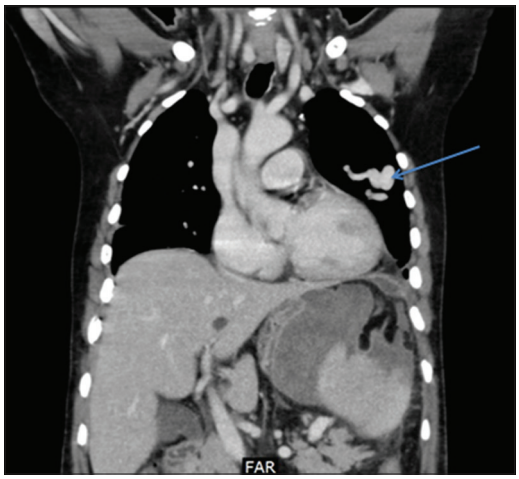

Fig2: Contrast-enhanced computerized tomography showing left upper lobe pulmonary AVM

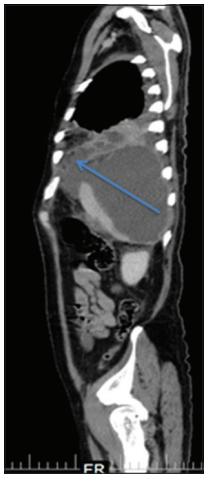

Figure 3: Contrast-enhanced computerized tomography (CT) showing a completely resolved splenic abscess but an increase in the size of the perisplenic hematoma as compared to previous CT, with a rent in left hemidiaphragm (blue arrow)

He was initiated on nasogastric tube feeds. 1 week later, repeat CT abdomen was done which showed resolving hematoma with decreasing pressure effects (Fig. 5). As he was clinically improving, antibiotics were stopped after the 5th week. Keeping in mind his clinical signs and pulmonary AVM, a diagnostic nasal endoscopy was done which showed multiple telangiectasias (Fig. 6), and hence, he was diagnosed as a case of Osler–Weber–Rendu disease as per curacao criteria.

The family members were advised to undergo screening. He was discharged in a stable condition, taking orally and with a plan to close the AVM through embolization after 2 weeks. He

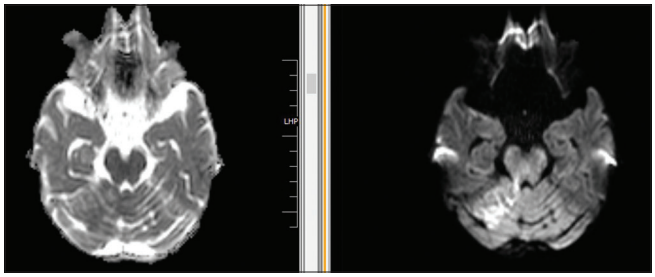

Figure 4: Acute infarct of the cerebellum on magnetic resonance imaging: T2-weighted image (left) and T1-weighted image (right)

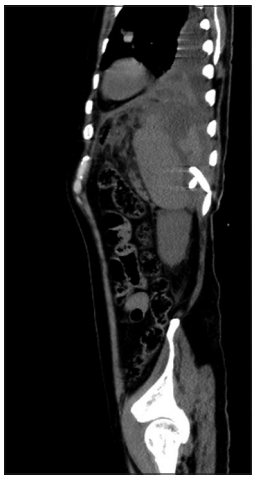

Figure 5: Contrast-enhanced computerized tomography showing resolving hematoma with decreased pressure effects

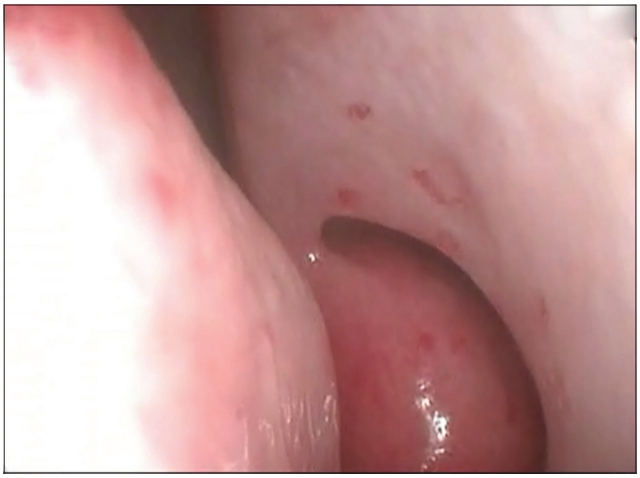

Figure 6: Diagnostic nasal endoscopy showing multiple telangiectasias

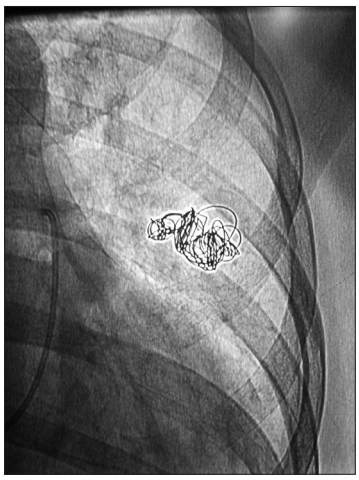

Figure 7: Radiological coil embolization done to close the pulmonary arteriovenous malformation

underwent successful radiological coil embolization following which his epistaxis resolved (Fig. 7).

DISCUSSION

HHT (Rendu–Osler–Weber syndrome) is an autosomal dominantly inherited disorder with a prevalence of 1 in 10000 to 1 in 5000 individuals [1]. It is believed that most, if not all, cases of HHT result from endoglin or anaplastic lymphoma kinase-1 haploinsufficiency (i.e., lack of sufficient protein for normal function), with the most consistent mechanism being the generation of a premature termination codon, resulting in non-sense-mediated decay of the mutant mRNA transcript [4]. Clinically, the organs most frequently affected by HHT are the lungs, brain, liver, and gastrointestinal tract [5]. Splenic involvement is considered a rare manifestation, with only a few case reports describing splenic involvement in patients with HHT [6-9].

Although splenic involvement has been described in HHT, splenic abscess as a complication of HHT has not been described so far. However, an isolated pulmonary AVM (PAVM) causing a splenic abscess along with a splenic infarction has been described [10]. Acute ischemic stroke is a common complication of HHT affecting ≤30% of individuals with PAVMs [2,11]. Stroke in the setting of HHT is usually secondary to aseptic thromboemboli passing through PAVMs [2]. Splenic infarcts and abscesses probably occur through the same mechanism of paradoxical embolism. Hence, this case is a rare manifestation where a splenic abscess and a perisplenic hematoma, along with multiple acute as well as chronic cerebrovascular accidents of both anterior and posterior circulation, of varying age, occurred in a single patient with HHT.

Early intervention for PAVM is important to prevent severe complications. Embolization of all angiographically visible PAVM prevents any central nervous system complications [2].

CONCLUSION

Splenic abscess and hematoma can occur as complications of HHT with a PAVM. Recurrent strokes can also occur in such a patient. The definitive management for the prevention of further similar instances would be closure of the PAVM and pre-emptive screening to look for AVMs in other sites of the body.

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