Title: Dieulafoy's disease of the bronchus: a possible mistake

Authors: Barisione E., Ravera S., Ferretti G.*, Salio M.;[1]

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Abstract

We present a case of a 57 year old woman who suffered from massive hemoptysis; she was sent to us for a suspect neoformant lesion. We assumed it might be Dielafoy's disease and proceeded with an imaging study that confirmed the diagnosis; after embolization, the patient no longer showed signs of bleeding. In brief, we concluded that whenever there is a suspect Dielafoy's disease, the biopsy has to be avoided.

Background

Dieulafoy's disease of the bronchus is supposedly very rare. In fact only few cases are reported in literature [4; 8]. This condition should be clinically suspected in heavy smokers with recurring and unexplained episodes of massive hemoptysis. The bleeding can occur immediately after the biopsy and/or after an interval of up to 12 days. The diagnosis can be made through imaging. Angiographic images document that this vascular malformation is based on a left-to-right shunt, with a bronchial artery draining into a pulmonary artery. Endobronchial ultrasound may be helpful in detecting the vascular nature of the lesion [1].

Case report

[1] Azienda Ospedaliera "SS.Antonio e Biagio e C.Arrigo" di Alessandria;

* Corresponding author
E-mail: gferretti@ospedale.al.it;

A 57 year old woman, non smoker, non atopic, was sent to us so we could study a neoformant lesion at the beginning of the superior right bronchus (Figure 1). She had been admitted to another hospital in February 2012 after suffering from seven episodes of massive hemoptysis. At the bronchoscopy there was no blood in the bronchial tree, but a little lesion with normal mucosa was present in the superior bronchus. The biopsy was followed by a massive hemoptysis episode that stopped only after 4 doses of tranxamic acid 5ml/500mg. During the emergency the patient had a hypotensive crisis, so only after the bleeding ceased she was transferred to the intensive care unit for monitoring of the hemodynamic functions. Finally, one hour later, a bronchoscopy was performed confirming the bleeding had stopped. In March a CT/PET was practiced and proved negative for hypercaptations. The histological evidence of the biopsy was normal bronchial mucosa with conserved structure, so this report was considered negative for neoplastic lesion. When she arrived at our hospital at the end of March, her doctors suggested a biopsy to be carried out with a rigid bronchoscope, which is safer in case of bleeding. However, after taking view of the histological description and visual image of the previous bronchoscopy we decided to use a flexible bronchoscopy in the presence of an MD anaesthesiologist. We found a lesion at the beginning of the medium bronchus (Figure 2); it was about 1-2 mm, raised from the surface with a white cap and covered form, apparently normal mucosa, but no lesion in the right upper bronchus, probably because this lesion had disappeared after the previous biopsy. Suspecting Dieulafoy's disease, we didn't carry out a biopsy of the lesion and proceeded to an x-ray study. The arteriography showed convoluted and ectatic bronchial vascular structures, particularly around and behind the trachea and around the right bronchus. (Figure 3). An embolization of the right bronchial artery and in particular of the common tract of the intercostal bronchial trunk was then performed using three 5mm spirals (Figure 4).

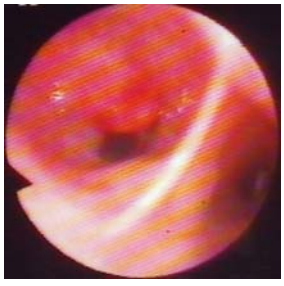

Figure 1: lesion at the beginning of superior right bronchus

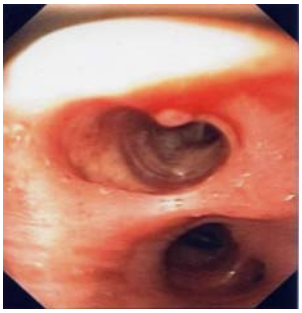

Figure 2: lesion at the beginning of medium bronchus

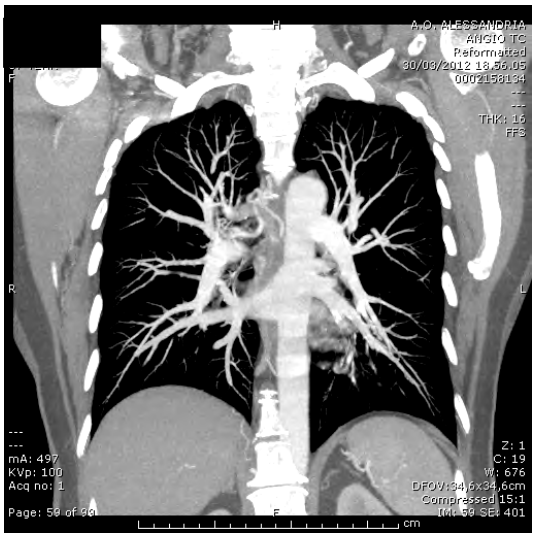

Figure 3: Arteriography

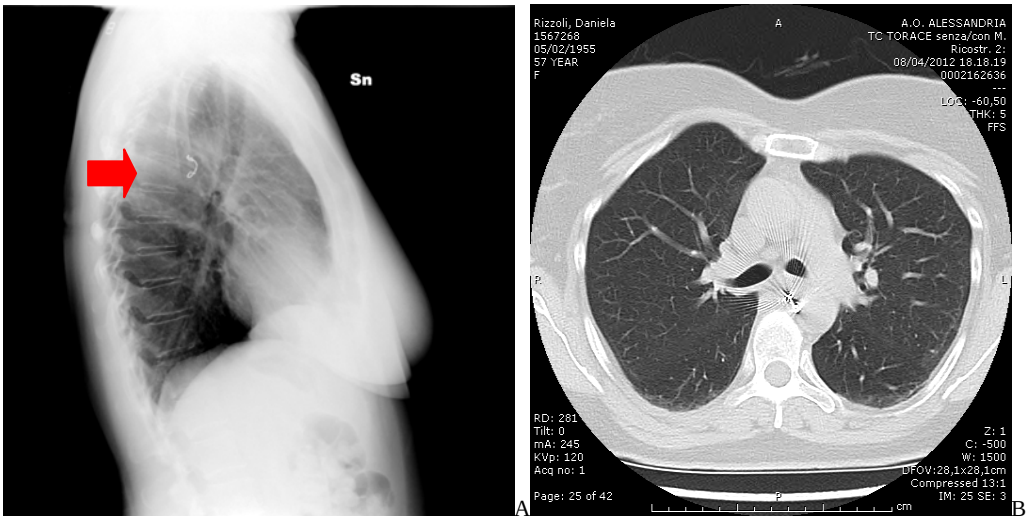

Figure 4

Spiral in the right bronchial artery, [A: X-ray B: CT]

Discussion

Dieulafoy's disease is an extremely rare vascular anomaly, characterized by the presence of a dysplastic artery in the sub-mucosa. At present, there are few proven cases reported in literature [2-3-4]. The pathogenesis of Dieulafoy disease remains unclear. It was first reported in the gastrointestinal tract [5]; more recently it has also been described in the respiratory tract [6-7]. While in the gastrointestinal tract the bleeding is often spontaneous but also fatal,

in the bronchial tree, profused bleeding often occurs after a biopsy. However, cases of spontaneous bleeding have also been described [3]. It is still unknown whether the origin of the anomaly is congenital or acquired, but age and/or tobacco use are thought to have an influence on the occurrence of the disease [2]. The trigger factor of the vessel rupture is unknown. Furthermore, the nature of the bleeding vessel remains controversial. Dieulafoy's disease of the bronchus is probably underestimated. Massive hemoptysis is a life threatening condition associated with a mortality rate exceeding 50% in the absence of adequate treatment [8-9]. The characteristics of the lesion are very non-specific, but in the presence of a small (usually < 1 cm), sessile, non pulsating nodular lesion, , often with a white cap, and apparently normal mucosa, Dieulafoy's disease should always be taken into consideration. The respiratory epithelium shows focal squamous metaplasia and diffused thickening of the basal membrane. In bronchial Dieulafoy's disease, selective embolization has been suggested as a method for stopping the bleeding [10-11] and only in few cases the patient required surgical resection[12].

Conclusion

In brief, Dieulafoy's disease of the bronchus, is more frequent than we think, so we have to consider the option when we have a patient with recurring massive hemoptysis, which cannot otherwise be explained. The biopsy, in this case, obviously has to be avoided, even when no active bleeding is evident.

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