Multiple aneurysms and Rosai-Dorfman disease: association or coincidence?

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DESCRIPTION

A 29-year-old man presented with a 1-month history of pain and numbness in his right arm and hypertension for 2 months, which was refractory to a combination of antihypertensive drugs. Physical examination revealed multiple pulsatile swellings in upper limbs and bilateral proptosis. No lymphadenopathy was noticed. The patient's blood pressure was 210/105 mm Hg. CT angiogram showed aneurysmal dilations in the left external carotid artery, left internal carotid artery, both axillary arteries and both renal arteries (figure 1). The right renal artery showed thrombosis (figure 2) and haemorrhage with non-functioning right kidney. Except for serum creatinine of 1.6 mg%, laboratory reports, including antinuclear antibody profile, c-ANCA (antineutrophil cytoplasmic antibody), p-ANCA, whole body positron emission tomography (PET) CT scan and thyroid profile, were normal. The patient had

Figure 2 Right renal artery showing thrombosis and haemorrhage, and aneurysm in the left renal artery.

Figure 1 CT angiogram showing aneurysmal dilations in left internal carotid artery, left external carotid artery, both axillary arteries and both renal arteries.

undergone surgical excision of a sinonasal mass 2 years earlier, the histopathological examination of which revealed Rosai-Dorfman disease (RDD).

RDD is a rare histiocytic disorder with excessive production of white cells called non-Langerhans sinus histiocytes.¹ These cells accumulate in various areas of the body, leading to malfunction. They mainly deposit in lymph nodes, particularly in the cervical region. The condition was first described as a distinct pathological entity by Rosai and Dorfman in 1969.² Involvement of extra nodal sites including orbits with proptosis, salivary glands, testis and upper respiratory tract, has been reported.³ There have been reports of unconfirmed vasculitis in RDD and we are reporting a case of multiple aneurysms in a patient with RDD. A definite association between RDD and multiple aneurysms still needs to be established.

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