Extraadrenal pheochromocytoma based in organ of Zuckerkandl.

Clinical History:

1 year history of arterial hypertension

Imaging Findings:

The patient presented with a 1 year history of arterial hypertension, headaches and migraine. The patient underwent CT, MIBG, venography and aortography.

Discussion:

Pheochromocytomas may present with vague symptoms due to excess catecholamine production: the majority present with paroxysmal (47%) or sustained (37%) hypertension, although other presentations include nausea, headaches or flushing. Urinary VMA (vanillyl mandelic acid) excretion is elevated in 54%. They may occur as part of a multiple endocrine neoplasia syndrome, or in association with a neuroectodermal disorder such as tuberous sclerosis or Von Hippel-Lindau syndrome.

Pheochromocytomas have been called the “10% tumour” since:
- 10% are located outside the adrenal glands (particularly in the para-aortic sympathetic chain or the organ of Zuckerkandl);
- 10% are malignant (although 40% of extra-adrenal tumours are);
- 10% are familial;
- 10% are bilateral or multiple.

Treatment is by surgical resection after pharmacological blockade. This patient was a Jehovah’s Witness and completely refused blood transfusion. Therefore pre-operative embolisation (with pharmacological blockade) was performed with PVA particles. This was successfully performed and allowed complete surgical resection with minimal blood loss at operation.

Differential Diagnosis List: Extraadrenal pheochromocytoma based in organ of Zuckerkandl.
**Final Diagnosis:** Extraadrenal pheochromocytoma based in organ of Zuckerkandl.

**References:**


**Description:** CT scan demonstrates a 5cm soft tissue retroperitoneal mass in the region of the lower abdomen. The mass demonstrates peripheral vascular enhancement with intravenous contrast in the arterial phase. **Origin:**
Description: MIBG scan demonstrates uptake in an area corresponding to the mass seen on CT.
Origin:
**Description:** Venography via a left femoral vein puncture demonstrates extrinsic compression of the left common iliac vein by a rounded mass. **Origin:**
Description: Aortography via a right common femoral artery approach demonstrates a complex vascular mass inferior to the origin of the inferior mesenteric artery, which has a prominent vascular blush and takes a feeding supply from several vessels in this region including the superior mesenteric artery. Origin:
Description: Aortography via a right common femoral artery approach demonstrates a complex vascular mass inferior to the origin of the inferior mesenteric artery, which has a prominent vascular blush and takes a feeding supply from several vessels in this region including the superior mesenteric artery. Origin:
Description: Post embolisation images demonstrate abolition of tumour vascularity. Surgery was subsequently performed with minimal blood loss. Origin: