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Acral Ischemia Secondary to Pheochromocytoma☆

Isquemia acral secundaria a feocromocitoma

Catecholamine-producing tumors are uncommon and called pheochromocytomas when their origin is intra-adrenal and paragangliomas when they are extra-adrenal.[1] The symptoms of pheochromocytoma include maintained or paroxysmal hypertension, headaches, palpitations and sweating. However, its symptoms can vary greatly, which is why it has been called "the great simulator".[1] Pheochromocytoma is rarely a cause of peripheral ischemia, but when this occurs it can lead to necrosis or gangrene, due in most cases to extreme vasoconstriction or diffuse arterial vasospasms induced by hypercatecholaminemia.[1–10] This vasospasm induced by catecholamines can easily be overlooked if the patient does not present other symptoms characteristic of pheochromocytoma; some patients may even have a history of intermittent claudication, a situation that can hinder and delay diagnosis.[1–10]

We report the case of a patient with a pheochromocytoma that presented multiple clinical manifestations and severe ischemic involvement of both the upper and lower limbs.

The patient is a 76-year-old woman with a history of hypertension under treatment for many years, ischemic stroke 20 years earlier with recovery and minimal sequelae, and gastric surgery for a bleeding ulcer 10 years ago. In the emergency room, she reported persistent intense headache for the previous 5 days associated with dizziness, nausea and vomiting. At this time, she was diagnosed with a hypertensive crisis, which subsided with medical treatment. She returned 24 h later and reported the same symptoms as before, in addition to epigastralgia radiating toward the thorax and diaphoresis. Upon examination, blood pressure was 185/90 mmHg, electrocardiogram showed a decrease in ST and negative T waves, and lab work-up detected elevated myocardial ischemia enzymes, so the patient was diagnosed with hypertensive crisis and acute coronary syndrome. 36 h after admission, the patient presented neurological deterioration with a Glasgow index of 12 and subsequent acute renal failure and respiratory failure, so she was transferred to the intensive care unit and intubated. Abdominal computed tomography (CT) scan showed a right adrenal mass measuring 8 cm in diameter; urine analysis demonstrated increased catecholamines (metanephrine 857 μg/24 h [Nr: 60–350]; noradrenaline 99 μg/24 h [Nr: 12–86]; adrenaline 45 μg/24 h [Nr: 2–23]). Blood pressure was controlled with iv labetalol during the 24 h of intubation, after which the treatment was changed to phenoxybenzamine and oral antihypertensive drugs. The patient presented an episode of atrial fibrillation that reverted to sinus rhythm with amiodarone. Renal failure and encephalopathy progressively improved.

During the hospital stay, the patient presented progressive acral ischemia of the 4 limbs, with necrosis of the fingers and toes (Fig. 1). Treatment was initiated with acetylsalicylic acid and perfusion of buflomedil. One month after admission to the hospital, right adrenalectomy was performed, initially by laparoscopy but then requiring conversion to subcostal laparotomy due adhesions caused by previous gastric surgery. The patient presented a favorable postoperative period, with progressive remission of the symptoms. The ischemic lesions of the 4 limbs became localized, and 53 days after admission, the patient required bilateral supramalleolar subcondylar amputation and metacarpophalangeal amputations of the fingers of both hands and interphalangeal amputations in the 2 thumbs. The patient recovered adequately. Vasculopathy and peripheral tissue ischemia are exceptional manifestations of pheochromocytoma and are due to the intense vasoconstriction of the cutaneous vessels in response to sympathetic overstimulation.[1,2] Early diagnosis of these cases is important to avoid vasoconstrictive medication, which may worsen the distal ischemia of both the upper and lower extremities.[3–8]

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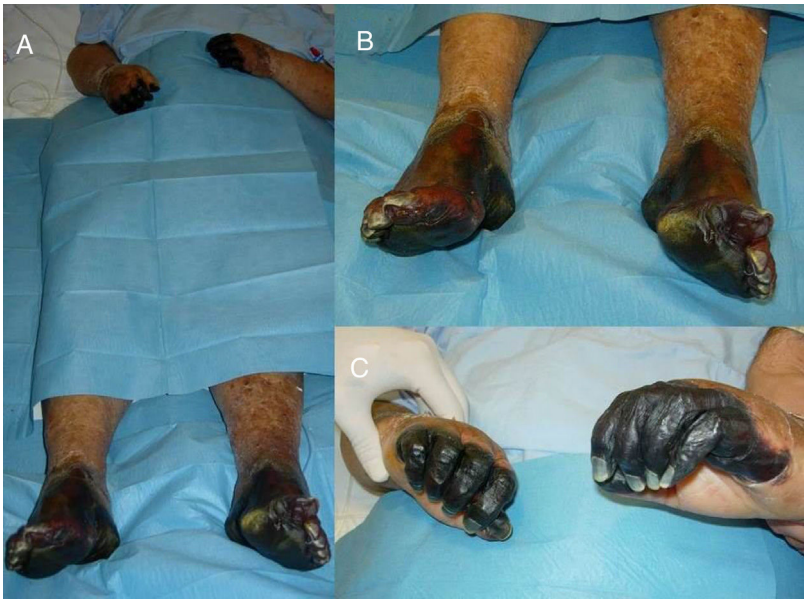

Fig. 1 – (A) Image of the patient during the acute episode of acral ischemia of both upper and lower extremities; (B) detailed image of the ischemia of the lower limbs; (C) detailed image of the ischemia of the upper limbs.

Table 1 – Cases of Pheochromocytomas With Distal Acral Involvement.

	Author	Age (yrs)	Sex	Size	Malignant Pheochromocytoma	Side	UE Involvement	LE Involvement	Adrenalectomy	LE Amputation	UE Amputation	Exitus
1	Rios (this study)	76	Female	8 cm	No	Right	Yes	Yes	Yes	Yes	Yes	No
2	Lutchmann et al.[1]	69	Female	5 cm	No	Left	Yes	Yes	Yes	Yes	Yes	No
3	Balbir-Gurman et al.[2]	63	Female	4.5 cm	No	Left	No	Right foot	Yes	No	No	No
4	Bessis et al.[3]	28	Male	6 cm	No	Left	No	Left foot	Yes	No	No	No
5	Tack y Lenders[4]	41	Female	5 cm	No	Left	No	Right foot	Yes	No	No	No
6	Muehrcke y Bliss[5]	48	Male	3 cm	No	Right	No	Left foot	Yes	Yes	No	No
7	Januszewicz y Wocial[6]	NS/NC	NS/NC	NS/NC	NS/NC	NS/NC	No	Yes	No	No	No	No
8	Borregana et al.[7]	37	Female	5 cm	No	Right	No	Right foot	No	No	No	Yes
9	Radtke et al.[8]	59	Female	5 cm	No	Left	No	Yes	Yes	No	No	No
10	Radtke et al.[8]	41	Female	12 cm	No	Left	No	Yes	Yes	No	No	No
11	Scharf et al.[9]	40	Female	1 kg (autopsy)	No	Right	No	Yes	No	No	No	Yes
12	Scharf et al.[9]	33	Female	–	Yes	Right	No	Yes	Yes	No	No	Yes
13	Engelman et al.[10]	63	Male	–	Yes	Right	Yes	Yes	No	No	No	No

LE: lower extremity; UE: upper extremity.

There is little experience worldwide in cases as dramatic as the one we present here, and fewer than 15 cases have been reported (Table 1). It is not a condition that exclusively affects older patients who present pheochromocytomas. Thus, almost half of case reports occur in patients under 45 years of age, as can be seen in Table 1. This indicates that it is more related with the production of catecholamines and the vasoconstrictor effect than the patient's baseline disease. It is observed that, in the majority of cases. acral ischemia mainly affects the lower limbs. However, in patients over the age of 60, it can also affect the upper limbs, as in our case.[1–10] The treatment in all cases is adrenalectomy, which is usually performed laparoscopically. Regarding the treatment of distal acral ischemia, it should be as conservative as possible so that the patient maintains as much personal autonomy as possible.[1–10] In conclusion, we can say that ischemia of distal acral areas, although rare, can occur in pheochromocytoma. Therefore, the diagnosis and early treatment of this disease is the way to avoid extensive amputations of the upper and lower limbs.

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ALPPS Tourniquet Complicated With Twisting of the Left Hepatic Vein. New Complications in New Hepatectomies☆

ALPPS torniquete complicado con giro de la vena suprahepática izquierda. Nuevas complicaciones en nuevas hepatectomías

Recently, revolutionary techniques have been described to avoid liver failure secondary to major hepatectomies.[1] Robles et al.[2] have described the associated liver tourniquet and right portal vein occlusion technique for staging hepatectomy (ALTPS) where portal ligation is performed and a tourniquet is placed over the parenchymal division line, accelerating hypertrophy and achieving effective regeneration in the first 7 days, with acceptable morbidity and mortality rates. The following is a case report of a complicated ALTPS with rotation of the left hepatic vein (HV). The patient is a 57-year-old woman with a history of laparoscopy-assisted sigmoidectomy in December 2014 for moderately differentiated adenocarcinoma of the sigmoid colon, pT3 N0. One year after surgery, elevated carcinoembryonic antigen (14.1 ng/mL) was detected, which had previously been normal (0–3 ng/mL). Computed tomography (CT) showed a solitary lung mass measuring 10 mm in the left upper lobe and 2 hepatic lesions, one in segment IV (18 mm) and another subcapsular in segment VII (8 mm), suggestive of metastasis. She received 2 cycles of chemotherapy with leucovorin, oxaliplatin and fluorouracil (FOLFOX), after which a new CT with vascular reconstruction and volumetry and a PET showed an increase in the size of the lesion in segment IV to 22 mm (no vena cava compromise) and stability of the hepatic lesion in segment VII and the lung lesion. The oncology committee discussed the case and decided to resect the liver lesions, followed by a second surgery to treat the lung lesion.

Intraoperative ultrasound confirmed the presence of the lesion located in segment VII; the lesion in segment IV was 4 cm and in close contact with the right and middle hepatic veins. The left suprahepatic vein was tumor-free. The residual liver volume (RLV) of the left lobe was 231 mL, representing 19% (total liver volume 1178 mL) for a patient weighing 77 kg. We decided to perform ALTPS following the volumetric criteria of Morales et al.[3] due to the need for rapid hypertrophy for the second operation and the treatment of the lung disease in the shortest time possible. The immediate postoperative period transpired without complications. On the tenth day, CT confirmed an RLV of 463 mL, which was 43%, and a second intervention was undertaken (Fig. 1a). A compromised vena cava was observed due to the segment IV lesion at the root of the right and middle hepatic veins. We then decided to resect

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