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Paraneoplastic Polyarteritis Nodosa Revealing an Ovarian Mass: Report of a Case and Review of the Literature

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Abstract **Case Report**

Polyarteritis nodosa (PAN) is a necrotizing vasculitis of medium and small arteries usually manifesting in a systemic form. In rare cases, PAN may reveal an underlying malignant disease. We report the case of a 62-year-old patient hospitalized for etiological assessment of chronic polyarthritis which revealed an ovarian mass associated with paraneoplastic polyarteritis nodosa.

Keywords: Polyarteritis Nodosa, Vasculitis, Paraneoplastic, Polyarthritis, Ovarian Mass.

INTRODUCTION

Polyarteritis nodosa (PAN) is a necrotizing vasculitis with a predilection for medium-sized arteries that can be localized to almost all organs. Numerous clinical manifestations are encountered in this condition depending on the location of the malignant tumor.

Paraneoplastic PAN is rare, which can reveal several cancers. Genital locations of PAN are uncommon [12]. We describe an observation of PAN revealing a malignant ovarian tumor.

CLINICAL CASE

We report the case of a patient aged 62, menopausal for 7 years. Ten months before her admission, the patient had presented inflammatory polyarthralgia small and large joints evolving in a context of apyrexia and deterioration of general condition made up of weight loss and asthenia. The patient did not report a history of fever, cough, diarrhea, or night sweats. The general examination finds a conscious patient, hemodynamically and respiratory stable, cachectic.

The osteoarticular examination reveals a polyarthritis of small and large joints. The clinical examination revealed a erythema nodosum on the legs and lower part of the thighs bilaterally (Figure 1) without lymphadenopathy or clinically detectable breast nodule.

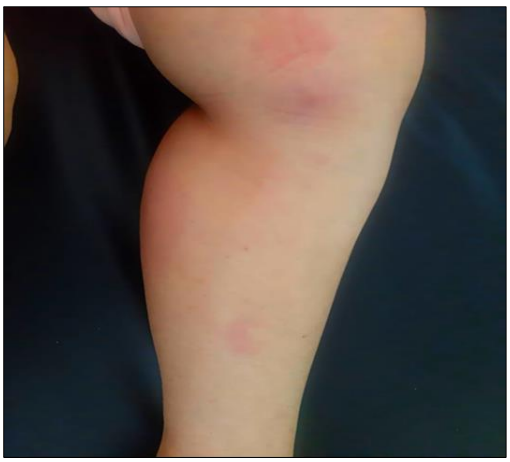

Figure 1: Erythema nodosum lesionpresent on the lower part of the thigh and the left leg

The assessment of polyarteritis nodosa carried out initially found anemia at 9.1 g/dlr associated with an isolated biological inflammatory syndrome with a CRP at 72 mg/L and an ESR (1st hour) at 89. The blood ionogram, renal function and liver tests are normal. Protein electrophoresis reveals an inflammatory syndrome with hypoalbuminemia, biclonal profile in gamma globulins with hypergammaglobulinemia. The immunological assessment made up of antinuclear antibodies, rheumatoid factor, anti-CCP and vasculitis assessment returned normal. The infectious investigation, the tuberculosis assessment as well as the converting enzyme and angiotensin were negative. Liver serologies were negative.

A skin biopsy was performed which revealed vasculitis mainly affecting medium-sized vessels with fibrinoid necrosis and cytosteatonecrosis lesion consistent with polyarteritis nodosa **(figure 2).**

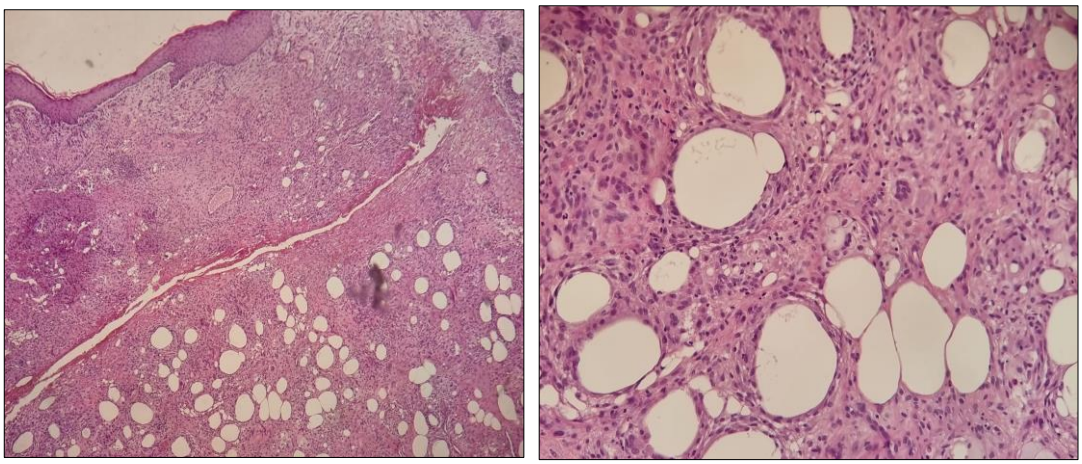

Figure 2: Skin biopsy

Radiologically, a CT chest revealed suspicious-looking bilateral nodules and micronodules of the pulmonary fields, bilateral breast infiltration more marked on the right associated with bilateral axillary and left supraclavicular lymphadenopathy. A mammogram has showed an appearance of mastitisdiffuse straight non-univocal character and brea jest biopsies had revealed a dystrophic breast parenchyma without specific or suspicious lesion.

The search for markers revealed the presence of CA125 at a level of 299.4 IU/ml, the other markers were negative. Our patient underwent a pelvic ultrasound revealinga peritoneal effusion of moderate abundance, a right ovary not visualized and the left ovary appears normal (Figure 3).

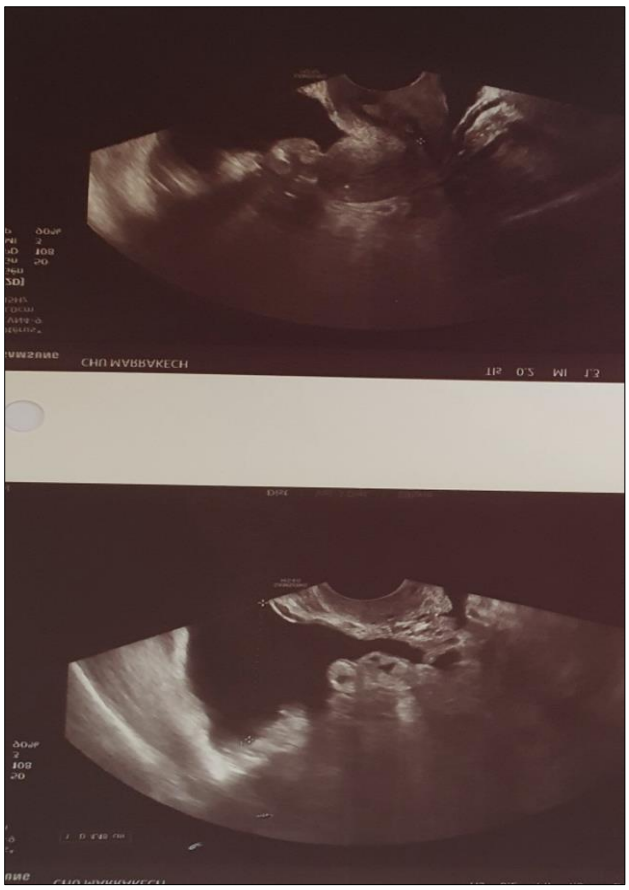

Figure 3: Endovaginal ultrasound showing a medium-sized peritoneal effusion

A pelvic MRI **(figure 4)** confirmed the presence of a suspicious-looking right lateral uterine mass, initially suggesting an ovarian origin associated with peritoneal carcinomatosis and pelvic and inguinal lymphadenopathy.

An exploratory laparotomy and bilateral adnexectomy are performed. Macroscopic and histological examination of the 2 ovaries revealed a parietal nodule, the site of a round cell tumor proliferation in the right ovary.

The diagnosis made in the patient was a paraneoplastic PAN revealing a malignant tumor of the right ovary.

The patient died before initiation of chemotherapy.

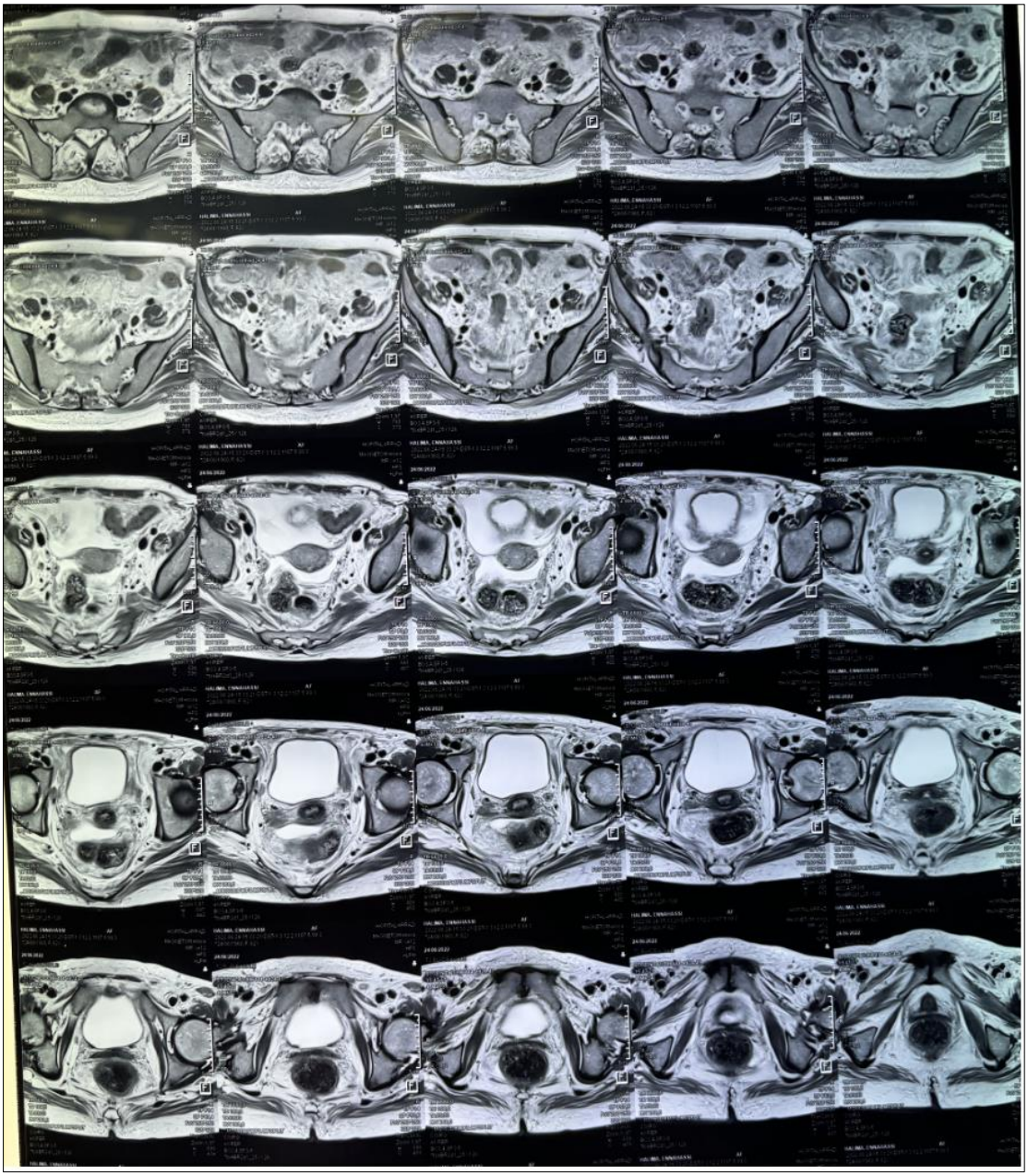

Figure 4: Alex

DISCUSSION

We report a case of paraneoplastic polyarteritis nodosa leading to the discovery of an ovarian tumor. PAN is rarely associated with malignant tumors and in this case, it is more often hematologic malignancies than solid neoplasia.

Polyarteritis nodosa (PAN) is a necrotizing arteritis of small and medium vessels that can affect any organ. It mainly affects middle-aged men and can have very varied clinical presentations. It has been demonstrated that systemic PAN is associated with hepatitis B while other infections (streptococci, parvovirus B19, HIV) cause cutaneous PAN [1]. In rare cases, PAN may reveal a malignancy.

Paraneoplastic PAN is uncommon, although its clinical presentation is similar to that of classic PAN. Its pathogenesis is still unclear, a cross-reaction between vascular endothelial cell antigens and primary malignancy antigens has been proposed [2].

A review of the literature found that PAN was associated with multiple myeloma [3], non-Hodgkin's lymphoma [3], leukemia [3], as well as solid tumors such as gastric cancer [4], colo -rectal [4], hypopharynx [5], lung [6], and bladder [7]. A diagnosis of PAN may be suspected based on the clinical presentation, but radiological or histological evidence is necessary to confirm the diagnosis when possible. The American College of Rheumatology has established 10 criteria for the classification of PAN in a patient with suspected vasculitis, and at least three or more criteria are necessary to make a diagnosis. These criteria have a specificity of 86.6% and a sensitivity of 82.2% [8].

A French study showed that these criteria are not always applicable and therefore histopathological evidence is useful to confirm the diagnosis [8]. A limited number of case reports involving vasculitis secondary to malignancy are reported in the literature. Our patient presented a very rare case of polyarteritis nodosa manifesting as an ovarian tumor. Although PAN may be an idiopathic diagnosis, the absence of previous symptoms and the atypical clinical presentation (chronic polyarthritis with deterioration of general condition) led to further investigations, which confirmed the diagnosis.

Thus, our patient's presentation suggests that malignancy may be a trigger for acute vasculitis and therefore, once malignancy is identified, therapy should be targeted at treating the malignancy rather than the vasculitis alone [10].

Although ovarian cancer has been described in relation to other paraneoplastic vasculitides such as Raynaud's phenomenon [11], its association with paraneoplastic polyarteritis nodosa has not been described in the literature.

Table 1: Clinical characteristics of cases of paraneoplastic polyarteritis nodosa in the literature

Authors	age	Sex	Clinical presentation	Associated signs	location	treatment	evolution	Reference
HAS.Grasland	81	F	Alteration of general condition	Long-term fever	uterine	Corticotherapy 1mg/kg/day	favorable	12
D. Veitch	65	H	Chest pain, left hand paresthesia, headache	Left hand hypoesthesia	cerebral	Corticosteroid therapy 70mg/day + cyclophosphamide 100mg/day Stop➔chemo-radiotherapy	favorable	13
Ahmed.U	54	H	Lower and upper limb pain	abdominal pain + MI edema	renal	Prednisone 80mg/d + cryoablation of the renal mass	favorable	14
Kolodziejczyk TC	78	F	Multiple erythematous to purplish macules on the lower limbs	-	Chronic myelogenous leukemia	Imatinib	favorable	15
Chircop.I	31	H	Acute ischemia of the left and right index finger	-	Cutaneous T-cell lymphoma	Gemcitabine	favorable	16
Our series	62	F	Chronic polyarthritis + deterioration of general condition	-	ovarian	-	deceased	-

CONCLUSION

Our patient represents the first documented case of paraneoplastic polyarteritis nodosa. The pathophysiology between systemic vasculitis and malignancy is still unclear requiring in-depth studies to elucidate this association. Thus, it is essential to seek the neoplastic origin of polyarteritis nodosa in the event of alarming signs such as the case of our patient.

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