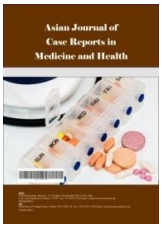

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A Lipomatous Axillary Mass Revealing an Eosinophilic Granulomatosis with Polyangiitis

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Authors' contributions

This work was carried out in collaboration among all authors. Authors RA and RT designed the study, wrote the protocol and wrote the first draft of the manuscript. Authors HT, IC and EC corrected the draft. Authors HT and MAS managed the literature searches. All authors read and approved the final manuscript.

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Case Study

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ABSTRACT

Eosinophilic granulomatosis with polyangiitis (EGPA), formerly called Churg-Strauss syndrome, is a condition characterized by asthma, high levels of eosinophils and vasculitis. It is characterized during its prodromal phase by an asthma of increasing severity. Vasculitis affects the lungs, the heart, the skin and the peripheral nerves. Skin involvement is common but may not be very suggestive.

The authors report the case of a 39-year-old man, with a medical history of asthma and sinusitis, who consulted in the ER for abdominal pain and partial occlusion syndrome. Diagnostic

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laparoscopy showed images of digestive vasculitis. Skin biopsy of an axillary subcutaneous mass which had lipomatous aspect was compatible with vasculitis associated with a perivascular infiltrate rich in eosinophils. Biology showed a persistent hyper eosinophilia in the blood. Antineutrophil cytoplasmic antibodies were negative. There was no evidence of pulmonary infiltrate or sinus opacity. The other cardiac, neurological and renal explorations were normal. The diagnosis of EGPA was suspected based on clinical and biological arguments. It was confirmed by skin biopsy of an atypical axillary mass. Treatment with corticosteroid therapy and immunosuppressants resulted in spectacular clinical improvement with a 3-year follow-up.
The cutaneous manifestations can have an essential contribution for the diagnosis of vasculitis. Early recognition of these lesions is essential for proper treatment and prevention of serious visceral complications.
Our observation is unique due to the atypical cutaneous manifestation which contributed to the early positive diagnosis of EGPA.

Keywords: Eosinophilic granulomatosis with polyangiitis; cutaneous manifestation; axillary mass.

1. INTRODUCTION

EGPA is a systemic and pulmonary vasculitis. Its etiopathogenesis is not yet elucidated. It is defined by its association with severe asthma and hyper eosinophilia in the blood and in the tissues [1]. Its high mortality makes it a serious systemic vasculitis [2].

Cutaneous vascular manifestations commonly occur during EGPA. They are polymorphic and non-specific but can reveal the disease in less than a quarter of the cases [3].

Vascular purpura, urticarial lesions and papules / nodules are the most frequent skin disorders and are observed in almost a third of the cases [3–5]. To our knowledge, a large soft mass simulating a lipoma has never been reported in the EGPA.

2. CASE PRESENTATION

Patient aged 39, with a history of moderate asthma from childhood treated with salbutamol when needed, as well as repeated sinusitis, consulted for acute abdominal pain with a partial occlusion syndrome.

The clinical examination showed a temperature at 37.8°C; blood pressure at 140 / 80mmHg; diffuse abdominal tenderness, as well as a soft, painless, lipomatous, 9x7x3 cm left axillary mass (Fig. 1).

Biology showed an inflammatory syndrome with a CRP at 51 mg / L and a sedimentation rate at 80, hyper eosinophilia at 7000 / mm3, normal renal function with negative 24-hour proteinuria. The search for polynuclear neutrophils (ANCA) antibodies was negative, as was the search for anti-nuclear factors and soluble nuclear antigen antibodies.

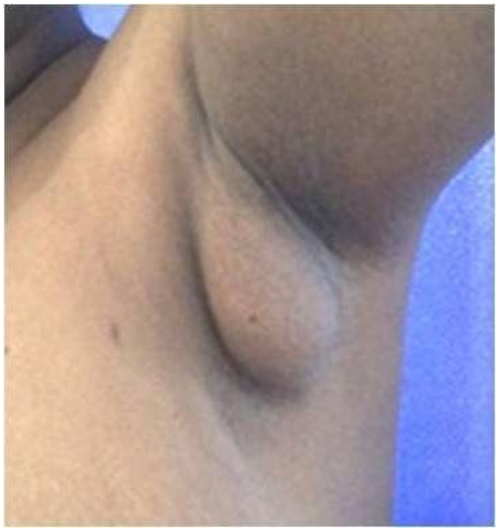

Fig. 1. Axillary mass

Upper gastrointestinal endoscopy showed grade B peptic esophagitis and congestive antral gastroraphy. The colonoscopy did not reveal any anomalies. A diagnostic laparoscopy was performed. The intraoperative aspect was in favor of vasculitis (Fig. 2). There were no signs of hemorrhage or perforation.

A surgical excision of the axillary mass whose histopathological study showed a fatty tissue of normal morphology (without arguments in favor of lipoma). There were fatty cells all around the tissue (Fig. 3).

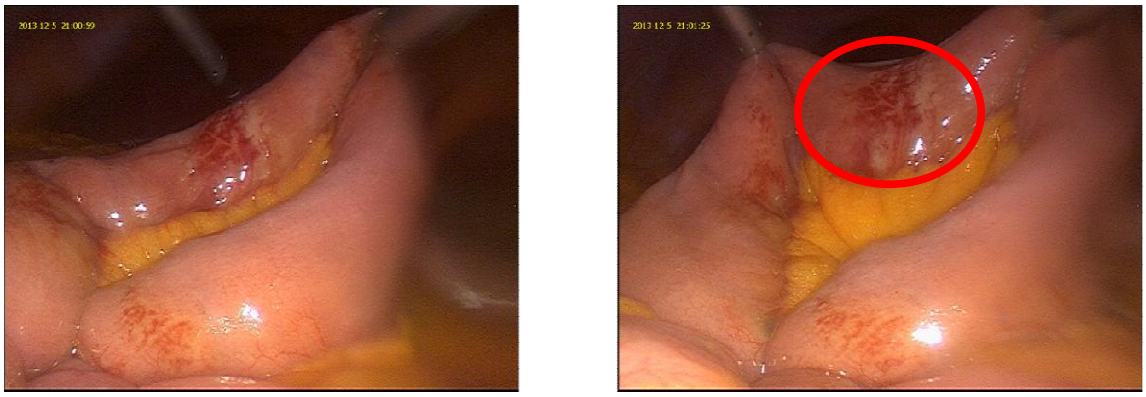

Fig. 2. Aspect of the laparoscopy was in favor of vasculitis

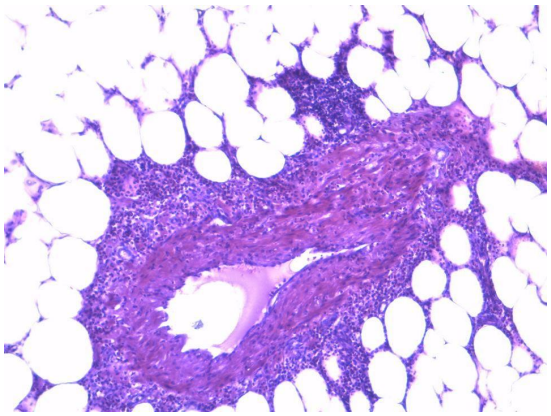

Fig. 3. Histopathological aspect showing fatty tissue

In addition, we noted on certain sectors, the presence around the vessels of medium caliber of an inflammatory infiltrate predominantly PNN and eosinophils. These dissociate the vascular wall at the level of the media and the adventitia (Fig. 4).

The ENT examination, the facial scanner, the electrocardiogram, the cardiac ultrasound, the chest scanner, the cerebral MRI and the electroneuromyogram were normal.

The diagnosis of EGPA was made, according to the criteria of the American College of Rheumatology, thanks to the history of asthma, sinusitis, hyper eosinophilia associated with an inflammation rich in eosinophile with necrotizing vasculitis of small and medium vessels.

Due to the digestive symptoms, three boluses of 500 mg / day of methylprednisolone IV were carried out, relayed by prednisone 1 mg / kg / day, associated with an induction treatment with cyclophosphamide in bolus prescribed at the dose of 0, 6 g / m2 on day 1, day 15 and day 30 then 3 additional boluses at a dose of 0.7 g / m2 every 21 days, followed by Azathiopirine.

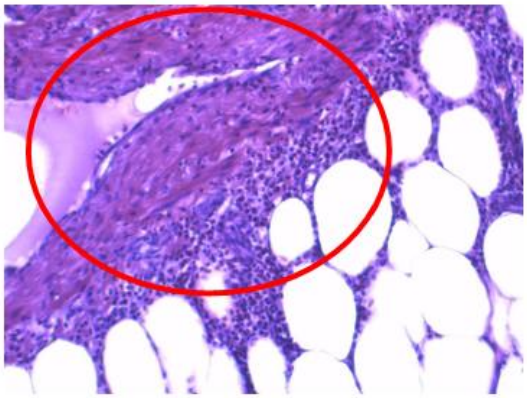

Fig. 4. Histopathological aspect: Inflammatory infiltrate of PNN and eosinophils

The evolution under treatment was spectacular with disappearance of abdominal pain. Since then, the patient has remained asymptomatic on long-term treatment with 10 mg / day of corticosteroids and 100 mg / day of Azathiopirine, with a 3-year follow-up. There was no recurrence of the mass.

3. DISCUSSION

EGPA can have multiple and varied clinical manifestations. Our patient had a multi-systemic EGPA including an atypical cutaneous involvement.

The diagnosis of EGPA is based on the identification of at least 4 of the 6 criteria of the classification defined by the American College of Rheumatology, with a sensitivity of 85% and a specificity of 99%. These criteria are: Asthma, blood eosinophilia> 10%, mono or polyneuropathy, labile pulmonary infiltrates, pain or sinus opacity and presence of extra-vascular eosinophils on a biopsy sample.

Severe late asthma is the most common revelation in EGPA [6]. However, other disorders can inaugurate the symptoms. Thus, the diagnosis might be overlooked. It is essential to have a rigorous diagnostic approach [7].

Making the diagnosis EGPA is problematic because it is difficult to obtain histopathological evidence [7]. In our case, we did not initially believe that the asymptomatic subcutaneous axillary mass was a cutaneous manifestation of EGPA [8]. Histopathological examination of this mass however revealed vasculitis with cutaneous eosinophilia, thus confirming our diagnosis.

Skin manifestations commonly occur during EGPA. A comprehensive literature review showed that skin manifestations occurred in 40 to 81% of EGPA patients and revealed the disease in 14% of patients [9].

Vascular purpura is the most common manifestation, affecting half of patients with skin involvement, followed by urticarial lesions, occurring in 12 to 31% of patients [10].

Other rarer skin manifestations can occur: Raynaud's syndrome, livedo reticularis, infiltrated nodules or papules, vesicles, skin rashes of Churg Strauss Syndrome or bubbles or even gangrene of a finger or toe [3,9].

The nodules are usually red or purplish, predominant on the finger, on the skull and on the extension face of the elbows or forearms. They are often bilateral and symmetrical.

However, the initial clinical presentation in our case with an isolated soft axillary mass was atypical. The differential diagnosis of an axillary mass is broad, including skin lesions, infections, haematoma, lymphadenopathy (hyperplastic, inflammatory, neoplastic or metastatic), accessory breast tissue, fibroadenoma, fibrocystic change, post-operative fluid collections, primary breast cancer and intramuscular neoplasms [11].

In the majority of the cases reported, the skin lesions allowed the histopathological diagnosis of EGPA. The main features are: vasculitis in 67%, eosinophilic infiltration in 58% and extravascular granuloma in 16% [12].

According to the diagnostic criteria of the EGPA, histopathological proof is necessary, and the skin constitutes a privileged site for biopsy.

In general, the prognosis has improved significantly since the use of corticoids and selected use of immunosuppressant agents for people with more severe disease. Systemic corticosteroids are still considered as the main treatment. Immunosuppressants are indicated in the presence of severe visceral involvement [13,14,15].

4. CONCLUSION

EGPA is a rare and severe systemic vasculitis, which can affect any organ. Pulmonary involvement occurs in almost all patients, and skin involvement is less common [14].

It is possible that non-specific skin manifestations, most often neglected or taken on account of other benign dermatological pathologies, may be useful in the early diagnosis of GEPA. If the disease is suspected, a skin biopsy must be systematically performed in the presence of any skin lesion. When treated with corticosteroids, the prognosis can change completely.

CONSENT

As per international standard or university standard, patient’s written consent has been collected and preserved by the author(s).

ETHICAL APPROVAL

As per international standard or university standard written ethical approval has been collected and preserved by the author(s).

COMPETING INTERESTS

Authors have declared that no competing interests exist.

REFERENCES

1. Churg J, Strauss L. Allergic granulomatosis, allergic angiitis, and

periarteritis nodosa. Am J Pathol. 1951;27(2):277-301.
2. Beltramo G, Cottenet J, Samson M, Foignot C, Malak CA, Quantin C, et al. Granulomatose éosinophilique avec polyangéite, éosinophilie pulmonaire et asthme: Une étude comparative épidémiologique de 5 ans utilisant la base de donnée PMSI. Revue des Maladies Respiratoires. 2017;34:A65.
3. Guillevin L, Cohen P, Gayraud M, Lhote F, Jarrousse B, Casassus P. Churg-Strauss syndrome. Clinical study and long-term follow-up of 96 patients. Medicine (Baltimore). 1999;78(1):26-37.
4. Lanham JG, Elkon KB, Pusey CD, Hughes GR. Systemic vasculitis with asthma and eosinophilia: A clinical approach to the Churg-Strauss syndrome. Medicine (Baltimore). 1984;63(2):65-81.
5. Finan MC, Winkelmann RK. The cutaneous extravascular necrotizing granuloma (Churg-Strauss granuloma) and systemic disease: A review of 27 cases. Medicine (Baltimore). 1983;62(3):142-58.
6. Bourg C, Morisset M, Lens V, Gerhard R, Feoli F, Wirtz G. Multiple ulcerated tracheobronchial mucosal lesions in the eosinophilic granulomatosis with polyangiitis. Rev Pneumol Clin. 2018; 74(6):516-519.
7. Zoubeidi H, Daoud F, Rachdi I, Somai M, Aydi Z, Ben Dhaou B, Boussema F. A perimyocarditis revealing eosinophilic granulomatosis with polyangiitis. Ann Cardiol Angeiol. 2020; 69(3):148-150
8. Kawakami T, Soma Y, Kawasaki K, Kawase A, Mizoguchi M. Initial cutaneous manifestations consistent with mononeuropathy multiplex in Churg-Strauss syndrome. Arch Dermatol. 2005;141(7):873-8.
9. Abe R, Hirayama T, Shimizu H. Disseminated subcutaneous nodules alone as manifestations of Churg-Strauss syndrome. Int J Dermatol. 2008;47(5): 532-3.
10. Davis MD, Daoud MS, McEvoy MT, Su WP. Cutaneous manifestations of Churg-Strauss syndrome: A clinicopathologic correlation. J Am Acad Dermatol. 1997;37(2 Pt 1):199-203.
11. Dialani V, James DF, Slanetz PJ. A practical approach to imaging the axilla. Insights Imaging. 2014;6(2):217-29.
12. Bosco L, Peroni A, Schena D, Colato C, Girolomoni G. Cutaneous manifestations of Churg-Strauss syndrome: Report of two cases and review of the literature. Clin Rheumatol. 2011;30(4):573-80.
13. Oh MJ, Lee JY, Kwon NH, Choi DC. Churg-Strauss syndrome: The clinical features and long-term follow-up of 17 patients. J Korean Med Sci. 2006;21(2): 265-71.
14. Solans R, Bosch JA, Pérez-Bocanegra C, Selva A, Huguet P, Alijotas J, et al. Churg-Strauss syndrome: Outcome and long-term follow-up of 32 patients. Rheumatology (Oxford). 2001;40(7):763-71.
15. Pagnoux C, Guilpain P, Guillevin L. Churg-Strauss syndrome. Curr Opin Rheumatol. 2007;19(1):25-32.

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