

Vasculitic neuropathy presenting as Churg-Strauss paraneoplastic syndrome

A rare association

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Churg-Strauss syndrome (CSS) is a rare necrotizing vasculitis, with incidence of 2.4 cases per million¹. It is composed of four of six criteria: asthma, eosinophilia greater than 10%, neuropathy, pulmonary infiltrate, sinus disease and a biopsy containing eosinophilic infiltrate².

Peripheral nerve system (PNS) compromise occurs around 75% of the cases and occasionally CSS may be of paraneoplastic origin^{3,4}.

We describe a rare case of paraneoplastic CSS associated with breast cancer, presented as severe peripheral polyneuropathy.

CASE

A 63-year-old mastectomized woman was admitted in October 2009 because of deep thrombosis. She had a triple negative breast neoplasia under control for five years and actually without medication. Two months before hospital admission, a severe sensory-motor axonal polyneuropathy developed and there was asthma initiated five years ago. During examination we observed deep areflexia in legs, weakness and atrophy of the interosseous muscles of hands and feet besides hypoesthesia in glove and stocking distribution. Laboratory tests revealed leukocytosis of 25,400 cells/mm³, with 57% eosinophils, LDH 707 U/L, creatinine 0.7 mg/dL, HSV 85 mm/h, positive rheumatoid factor 1:100, FAN 1:80, p-ANCA +, CA 15-313 U/mL and CEA 1, 4 ng/mL.

Brain MRI, chest and abdominal CT imaging were normal at hospital admission.

Biopsy of the sural nerve showed vasculitis with intense eosinophilic infiltration (Figure A and B).

During treatment with oral prednisone 60 mg and anticoagulants, thrombotic phenomenon recurred and there was worsening of neuropathy. Mensal pulse with 1gr cyclophosphamide was indicated during six months. Her condition worsened and after two months she died due metastases in the liver and lungs detected by CT.

DISCUSSION

Paraneoplastic syndrome refers to signs and symptoms caused by a neoplasm, but unrelated to direct tumor growth. They could appear before, after or concomitantly with the neoplasia, but around 71% of the cases occur before tumor discover or during recurrence⁵.

Its pathogenesis is due to secretions of substances like hormones or by immunomediated mechanisms resulting in neurological, rheumatological or vasculitic manifestation⁵. Angiitis associated with neoplasia is more commonly found in oncological-hematological disorders, but solid tumors of the lungs, breasts, prostate, colon and kidneys have been described^{3,4}.

CSS associated with cancer has been associated with non-Hodgkin B-cell lymphoma and melanoma². It is vasculitic involvement could also affect veins in active phase and although such event did not form the diagnostic criteria, they need to be kept in mind given that they are highly prevalent.

Acute neuropathy may simulate Guillain-Barré syndrome, but a subacute course of multiple mononeuropathy develops in 71% of the cases, with greatest impairment of the peroneal nerves. In the final stages, the

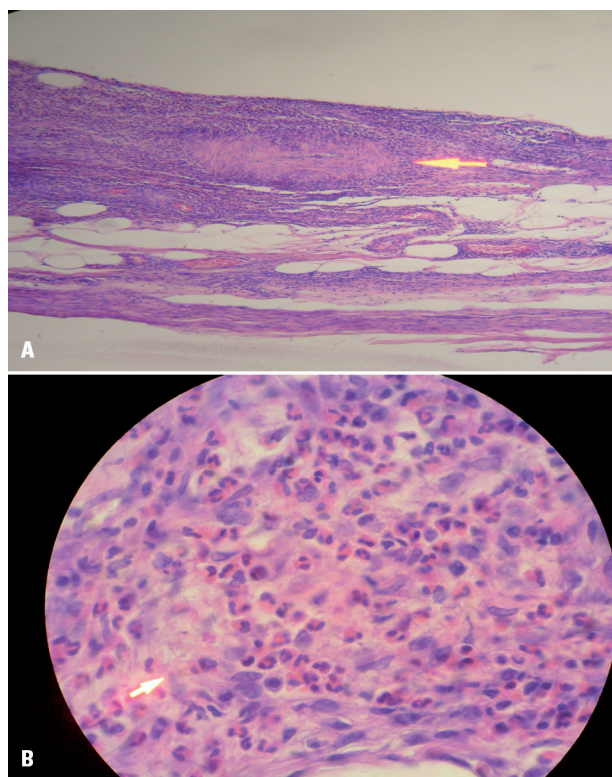


Figure. [A] Intense arteritis in peripheral nerve sheath (HE 40×). [B] Inflammatory infiltration with intense eosinophils around vessels producing vasculitic aspect (HE 400×).

patterns may converge in symmetrical axonal polyneuropathy in 29% of the cases. Central nervous system involvement is less common and ranges from 6% to 39%¹.

CSS as a paraneoplastic syndrome represents a rare condition and this association in a breast cancer patient becomes important to recognize since there are no reports of this conjunction.

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NEUROPATIA VASCULÍTICA COMO MANIFESTAÇÃO DE SÍNDROME DE CHURG-STRAUSS PARANEOPLÁSICA: UMA RARA ASSOCIAÇÃO

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