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Case Report

Dermatomyositis Related to a Male Germ Cell Carcinoma: a Case Report

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Abstract

Dermatomyositis is an autoimmune disorder that affects mainly muscles and skin. In some cases it may be associated to neoplasms occurring before the diagnosis of cancer. Its relation to germ cell tumor is not well documented, so we describe a case of a patient with history of muscle weakness and cutaneous rash that preceded cancer for 6 months. Dermatomyositis must remind of a possible occult neoplasm and its remission may follow the treatment of the tumor.

Keywords: Dermatomyositis. Germ cell. Skin. Neoplasms.

Introduction

Dermatomyositis is an idiopathic inflammatory myopathy of unknown cause. Its association with malignancy is documented, but no clear mechanism was identified. Up to one-fourth of patients with dermatomyositis may have an occult cancer.1 Although this association is recognized, there are few reports of dermatomyositis associated with germ cell cancer.2 Some data show a higher mortality by cancer in patients with dermatomyositis.1 We report a case of a male patient with dermatomyositis and germ cell cancer that had a successful resolution of dermatomyositis with chemotherapy for germ cell cancer.

Case Report

A 31 year old male patient with cutaneous rash and bilateral proximal muscle weakness of lower extremities and loss of weight presented to our center. Physical examination revealed motor loss with difficulty walking and slight skin rash in trunk and upper limbs, but otherwise unremarkable. Initial diagnostic evaluation with laboratory tests showed antinuclear antibody titer 1/320 (speckled pattern), elevated lactate dehydrogenase (DLH 1078 U/L), creatinine phosphokinase (4515U/L), and sedimentation rate (27mm). To test the hypothesis of dermatomyositis, an eletromiography was performed and revealed sensorimotor axonal polyneuropathy. Biopsy of quadriceps muscle showed interfascicular atrophy and lymphocytic inflammatory infiltrate. These factors, together with the clinical findings, are compatible with dermatomyositis.

He started steroids and methotrexate with improvement of symptoms. After 5 months, he complained of abdominal pain and leg weakness. Due to abdominal pain, we did an abdominal computed tomography (CT) that revealed a retroperitoneal mass (Figure 1A) involving

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bone and nerve roots. Chest CT showed multiple pulmonary nodes and mediastinal mass (Figure 2A).

In addition, a biopsy of retroperitoneal mass revealed undifferentiated neoplasm with immunohistochemical markers representing germ cell malignancy (positive strain for placental alkaline phosphatase-PLAP and alpha-fetoprotein), a typical seminoma. Ultrasonography of scrotum showed no mass. Marker study serum levels for germ cell tumor showed elevated alpha-fetoprotein (129,5U/L), DLH (3189U/L) and human chorionic gonadotropin (6.003 U/L). Although the pathological analysis showed a seminoma, he had an elevated alpha-fetoprotein, and it was considered as a non-seminomatous germ cell tumor, clinical stage IIIC, intermediate risk, and he started standard chemotherapy with PEB regimen (cisplatin 20mg/m² days 1-5, etoposide 100mg/m² days 1-5 and bleomycin 30 units days 1, 8 and 15), every 21 days. After first cycle, the patient recovered from muscle weakness and tumor markers achieved normal levels. Assessment with CT scan after third cycle revealed complete response in mediastinum and lungs (Figure 2B). He was kept on chemotherapy until sixth cycle and after that a residual retroperitoneal mass remained (Figure 1B). Although it was a non-seminomatous germ cell tumor, a PET-CT was done and it was unremarkable. A choice for no further therapy was done. During follow up, the residual mass grew, and the patient was submitted to surgery. Pathological specimen showed fibrosclerotic nodes with necrosis and calcified areas, as well as lymphoid aggregates and macrophages. No viable neoplastic cell was found. The patient has been on follow up without evidence of tumor, and no skin or muscle manifestation of dermatomyositis for 20 months after the end of chemotherapy.

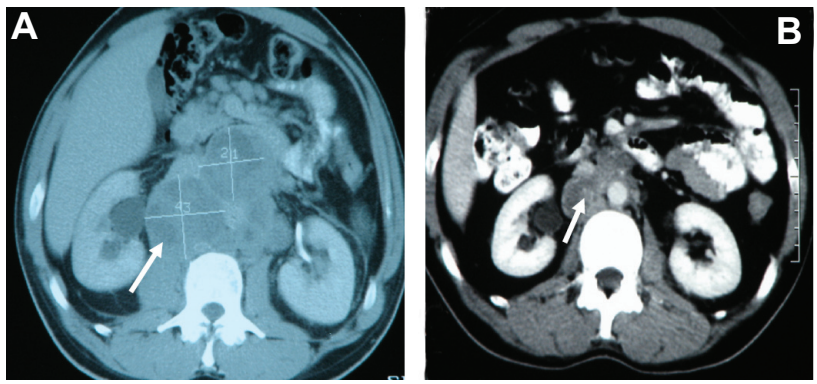

Figure 1 - Abdominal CT at the beginning (A) and after (B) of chemotherapy with residual retroperitoneal

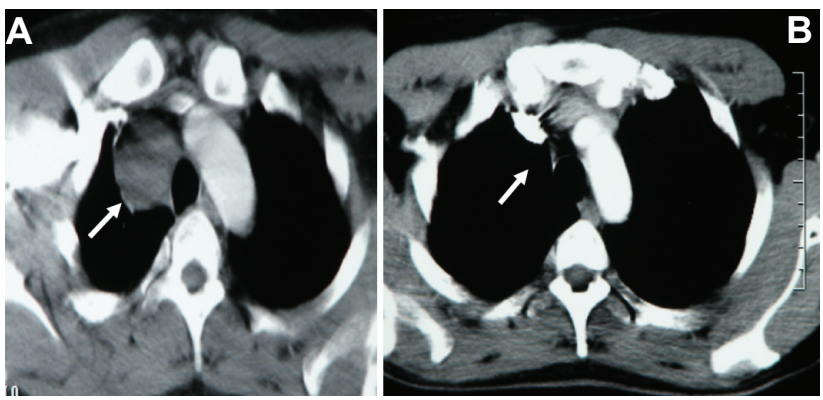

Figure 2 - Chest CT at diagnosis, showing mediastinal tumor (A), and after three cycles of chemotherapy (B)

Discussion

Many data in the medical literature reported frequent association of dermatomyositis and polymyositis with cancer3. Dermatomyositis is a paraneoplastic disorder of connective tissue and the process that explain increased risk of neoplasm is not defined. As it is an autoimmune disease, there are high levels of antibodies both to normal tissue and cancer cells.

A retrospective analysis of 788 patients with dermatomyositis or polymyositis showed a cancer incidence in this population of 9%. Cancer mortality was also higher than in the general population. 1 The most common neoplasm are lung, gastric, colorectal and ovarian, and less than ten cases of germ cell cancer and dermatomyositis is reported in the literature2,4-5. Our patient, as it happened in the other documented similar cases, first presented dermatomyositis and only some months later a tumor was found. The treatment of dermatomyositis, considering it as a paraneoplastic manifestation of cancer, is the standard management of the tumor. In our case, as it was a germ cell tumor, treatment was chemotherapy based in cisplatin and the patient had a good response of tumor and resolution of dermatomyositis6-8. In non-seminomatous tumor, PET-CT has limited predictive value, so all post-chemotherapy residual mass should be ressected, but in this case surgery was done later. However, there was no residual neoplasia9.

Conclusion

In summary the diagnosis of dermatomyositis is a key for search malignancy. Even when the tumor is not found at the onset of dermatomyositis, it is important to investigate the most frequent tumors for each age and gender and keep in mind that in young male with abdominal mass the testis should be examined.

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