Scleromyxedema: Possible association with seminoma


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Although scleromyxedema has been associated with neoplasm in rare instances, the literature showed no evidence of association with seminoma. We report a 43-year-old man who presented with a scleromyxedema and relapsed seminoma. The skin lesions of scleromyxedema cleared completely on treatment of seminoma with chemotherapy. (J Am Acad Dermatol 2000;42:875-8.)

Scleromyxedema is a rare fibromucinous disorder first described by Gottron in 1954. It is characterized by the presence of infiltrative skin lesions with the deposition of mucinous material in the papillary dermis. Scleromyxedema tends to have a chronic and progressive course with little tendency toward spontaneous remission.

The disease primarily involves the skin, although extracutaneous manifestations have been described. The most widely known association is a monoclonal gammopathy, usually of the IgG or IgA class. Other disorders associated with scleromyxedema are multiple myeloma, amyloidosis, various neurological, cardiovascular, and renal disorders. Collagen vascular and some dermatologic disorders have also been reported. A review of the literature suggests a rare association of scleromyxedema and malignancies, but no reports showed an association with seminoma.

In this report, we describe a case of a relapsed seminoma and scleromyxedema.

CASE REPORT

A 43-year-old man was diagnosed in 1991 to have right seminoma metastatic to para-aortic nodes. He was treated in Egypt with high inguinal orchiectomy, followed by radiation therapy to the para-aortic lymph nodes and pelvis, with a total of 3000 cGy. He remained healthy until October 1993, when he presented to King Faisal Specialist Hospital and Research Centre with a dry cough, shortness of breath, large mediastinal mass, and fullness in the left supraclavicular area. In addition to his respiratory symptoms, the patient also observed progressive thickening of the skin over the scalp, neck, upper trunk, and arms. A skin examination of these areas revealed indurated yellowish and violaceous papules, nodules, and plaques (Fig 1, A and B).
A skin biopsy specimen stained with alcian blue revealed dermal mucin (Fig 2, A and B).

The presence of satellite fibroblasts suggested the local production of mucin. A diagnosis of scleromyxedema was made.

A computed tomographic scan of the chest confirmed a huge, homogenous mass in the dorsal mediastinum, dislocating the large vessels, trachea, and esophagus anteriorly.

A computed tomographic-guided true-cut needle biopsy specimen of the mediastinal lymph node confirmed the diagnosis of recurrent seminoma. As part of the workup, serum protein electrophoresis was performed and showed normal values. After 2 cycles of chemotherapy, which was directed to the relapsed seminoma and included cisplatin, etoposide, and bleomycin, the skin lesions regressed in size (Fig 3) to clear completely after the end of the fourth cycle.
Scleromyxedema is a rare type of papular mucinosis characterized by diffuse sclerosis and a generalized lichenoid eruption. The dorsal region of the hands and fingers, the extensor area of the arms, legs, and face are typically involved. However, large areas of the body may be involved and diffuse induration, edema, and erythema may be present. The disease typically affects adults between the ages of 30 and 70 years, has no sex predilection, is usually chronic, and therapy is difficult.²³

Scleromyxedema primarily involves the skin, but systemic manifestations have been described.⁴¹² The most widely known association is a monoclonal gammopathy, usually of the IgG and mainly of lambda light chains.¹³¹⁴

An association of the disease with neoplasms has rarely been described in the literature⁵¹¹ (Table I).

**DISCUSSION**

Several weeks after chemotherapy, the patient developed sensory neuropathy, confirmed by a nerve conduction study, affecting both the median and ulnar nerves. He was followed up for 2.5 years with no recurrence in the skin lesions or any evidence of his original neoplasm.

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Table I. Reported neoplasms associated with scleromyxedema

<table>
<thead>
<tr>
<th>Number</th>
<th>Type of neoplasm</th>
<th>Reference</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Adenocarcinoma of the stomach</td>
<td>[5]</td>
</tr>
<tr>
<td>2</td>
<td>Carcinoma of the pancreas</td>
<td>[5]</td>
</tr>
<tr>
<td>3</td>
<td>Mucinous ovarian carcinoma</td>
<td>[6]</td>
</tr>
<tr>
<td>4</td>
<td>Squamous cell carcinoma of the bladder</td>
<td>[7]</td>
</tr>
<tr>
<td>5</td>
<td>Squamous cell carcinoma of the lip</td>
<td>[8]</td>
</tr>
<tr>
<td>6</td>
<td>Esophageal carcinoma</td>
<td>[9]</td>
</tr>
<tr>
<td>7</td>
<td>Hodgkin’s disease</td>
<td>[10]</td>
</tr>
<tr>
<td>9</td>
<td>Seminoma</td>
<td>Present case</td>
</tr>
</tbody>
</table>
None of them was associated with testicular seminoma. In our case, a scleromyxedema appeared at the time of seminoma relapse and cleared after chemotherapy directed to seminoma.

The treatment of scleromyxedema is notoriously difficult.\(^2\)\(^{15}\) Many chemotherapeutic agents such as melphalan, cyclophosphamide mechlorethamine hydrochloride, and methotrexate \(^2\)\(^3\)\(^{15}\)\(^{18}\) have been used; however, melphalan showed a dramatic effect with the softening of the skin, decreased cell proliferation, and reduced levels of paraprotein. Conversely, this effect was often temporary and the disease has proven fatal in 10 of 17 patients treated with this alkylating agent, predominantly from hematologic malignancies and septic complications.\(^2\)

Albert et al\(^{10}\) reported a 41-year-old woman with scleromyxedema, who was treated for 6 years with cytostatic drugs. During that time, the skin lesions followed a fluctuating but progressive course. After 6 years she developed Hodgkin’s lymphoma of the mixed cellularity type. Intensive cytostatic treatment (mustine, vincristine, procarbazine, prednisone, doxorubicin, vinblastine, and bleomycin) given for Hodgkin’s disease resulted in virtually complete disappearance of the scleromyxedema lesions.

Garcia et al\(^{11}\) reported a case of scleromyxedema with non-Hodgkin’s lymphoma, and combination chemotherapy with methotrexate, doxorubicin, cyclophosphamide, vincristine, prednisone, and bleomycin (MA COP-B) was given. The patient improved and cutaneous plaques and enlarged lymphatic nodes disappeared.

Although several chemotherapeutic agents have been used to treat scleromyxedema, no report shows the effect of cisplatin or etoposide, which were used in our case. However, bleomycin has been used in the treatment of 2 patients.\(^{10}\)\(^{11}\)

The rationale behind the use of chemotherapeutic agents is the proposed relationship of scleromyxedema to a plasma cell dyscrasia, which is thought to account for the monoclonal gammopathy.\(^2\) The sensory neuropathy of our case was suspected to be related to cisplatin. However, scleromyxedema has also been known to be associated with peripheral neuropathy.\(^{19}\)

In this case, we wonder if this dramatic response, with long-term remission of more than 2.5 years, is due to chemotherapy or related to the remission of the original disease. After 2.5 years of remission, the patient returned to his country.

We would like to emphasize the importance of a thorough clinical examination, including hidden areas such as the testis, as part of any evaluation of patients with scleromyxedema. We believe the recurrence of seminoma accompanied by scleromyxedema, and a parallel course of both conditions (ie, scleromyxedema cleared on remission of seminoma after chemotherapy directed toward seminoma) probably makes our patient a case of cutaneous paraneoplasia.\(^{20}\)

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REFERENCES


