Case report

Pulmonary infiltration from retroperitoneal carcinoma requiring diaphragm, chest wall and lung resection after thoracoabdominal access

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Received 10 January 2003; received in revised form 16 March 2003; accepted 21 March 2003

Abstract

Primary retroperitoneal carcinoma is a rare but highly aggressive tumour often resulting in infiltration of abdominal organs and diaphragm. We describe a case of retroperitoneal carcinoma infiltrating the diaphragm, the lower lobe of the left lung and ribs IX and X, but sparing the abdominal organs. After thoracoabdominal access, our patient underwent resection of the retroperitoneal carcinoma, left hemidiaphragm and ribs IX and X with wedge resection of the left lower lobe. Haematogenous lung metastasis from retroperitoneal carcinoma is well known; on the contrary, direct transdiaphragmatic lung invasion is very rare and requires thoracoabdominal access for a one step operation, representing one of the most stimulating challenges for thoracic surgeons.

1. Introduction

Primary retroperitoneal tumours (PRT) arise in the retroperitoneal space but are unrelated to the organs of this space. They are extremely rare, accounting for 0.05–0.2% of all solid neoplasms. They affect men and women alike with a peak incidence between 40 and 60 years of age [1]. Retroperitoneal carcinoma (adenocarcinoma, squamous, undifferentiated) is one of the rarest retroperitoneal tumours, accounting for 8% of all retroperitoneal lesions [2].

Surgery is the therapy of choice requiring a transperitoneal approach after median laparotomy or thoracoabdominal access. Radical resection is the most important factor influencing the prognosis; adjuvant radiochemotherapy is controversial as it does not seem to improve long-term survival [1–3].

2. Case report

A 54-year-old man was admitted to our department for a left subdiaphragmatic mass disclosed by chest CT scan (Fig. 1A).

The patient had been febrile for 2 months. Blood tests on admission showed haemoglobin (Hb): 7.2 g/dl, white blood count (WBC): 48,500 µl, platelets (PLT): 655,000, creatinine: 1.4 mg/dl. All other tests were normal. A blood cytosmear and blood analysis excluded haematologic diseases. Abdominal CT scan disclosed a left posterior subdiaphragmatic mass, in contact anteriad with the spleen and mediad with the left kidney. The lesion was colligate with solid central zones, strong contrast enhancement and a diameter of 5 × 6 cm. Pet scan showed inhomogeneous contrast uptake in the mass and intense medullary skeletal activation.

CT-guided fine needle aspiration biopsy showed necrotic cells weakly positive for HNF 116 (wide spectrum cytokeratin) and negative for CD 45; immunomorphological analysis was suspect for carcinoma. Preoperatively the patient received a blood transfusion (600 cc); post transfusional tests showed Hb: 8.8, WBC: 50,500 PLT: 579,000. The patient underwent thoracoabdominal access: no abdominal organ was infiltrated by the mass, whereas we observed diaphragm, chest wall and lung infiltration. The retroperitoneal carcinoma was resected together with part of the left hemidiaphragm, IX and X ribs and left lung lower lobe parenchyma by wedge resection. Goretex was used for chest wall reconstruction but no prosthesis was necessary to close the left hemidiaphragm aperture (Figs. 1B and 2).

Histological examination showed scarcely differentiated carcinoma with a high mitotic index, giant and fusiform cells infiltrating the diaphragm and lung; no rib
infiltration was observed. The immunohistochemical algorithm was: smooth muscle actin: focally positive; keratin: positive; vimentin: positive. Global immunomorphological examination disclosed a high mitotic index sarcomatoid carcinoma (soft tissue carcinoma – sarcoma).

During the post operative period, leucocytosis gradually decreased and haemoglobin stabilized at 12.5 g/dl. The patient was discharged 11 days after surgery in good general condition. The patient underwent radiotherapy and 11 months after surgery he is still alive.

3. Discussion

PRT arise from the retroperitoneal space, delimited at the back by the quadratus lumborum, iliopsoas, prevertebral muscles and vertebral bodies and anteriad by the posterior parietal peritoneum with the diaphragm in the upper part and the pelvic floor in the lower part.

PRT form a heterogeneous group of malignant mesenchymal and neuroectodermal neoplasms, accounting for 0.05–0.2% of all solid neoplasms. They mainly comprise sarcomatous lesions like liposarcoma (the most frequent), fibrosarcoma, malignant fibrous histiocytoma, leiomyosarcoma and rhabdomyosarcoma, whereas carcinoma and germ cell tumours are less common [2].

The most frequent symptoms are: palpable tumescence, pain, weight loss, fever, muscle weakness and anaemia; urinary, digestive and neurological symptoms are rare and tardy, due to venous stasis; PRT rarely cause paraneoplastic endocrine syndromes like hypoglycaemic syndrome [1]. CT scan and ultrasound give the most information on mass structure (solid, cystic, mixed) and relations with other structures.

Surgery is the therapy of choice for PRT. Radical resection, however, is possible only in half the cases (33–70%) and often entails resection of adjacent organs (kidney, pancreas, colon and, as in our case, ribs, diaphragm and lung); unresectable tumours can be clipped to help plan radiotherapy. Either a long vertical incision or bilateral transverse incision can be used to explore a retroperitoneal mass; some Authors [2] discourage lateral positioning and thoracoabdominal or flank incision because they do not provide optimum access to the midline, where dissection around vascular structures is often critical; in this case, instead, we performed a classic thoracoabdominal access (thoraco-freno-laparotomy) providing radical resection. Operative mortality ranges from 2 to 21%.

Recurrence is common even after radical resection (20–87%) with a medium disease-free interval of 20 months. Recurrence frequently occurs because the pseudocapsule circumscribing the tumour only results from compression of surrounding organs and local inflammation and does not prevent neoplastic cells spreading. The overall 5 year
survival rate in resected patients is 15–50%, but it is 32–74% in patients who have undergone radical resection.

There is a need for research in chemotherapy and radiotherapy of retroperitoneal malignant neoplasms; some Authors feel there is value in combination radiochemotherapy; it appears that for radiotherapy to be effective, relatively high doses (40 Gy) are required [2]. Polychemotherapy is mainly represented by PVB (cis-platinum, vinblastine, bleomycin); PVA (cis-platinum, etoposide, adriamycin); VLB (vinblastine); CAc (cyclophosphamide, actinomycin D).

Adjuvant therapies offer little benefit although some authors observed a slightly reduced recurrence rate in patients receiving post-operative radio and/or chemotherapy [4,5].

Aggressive surgery is the most important factor in the treatment of retroperitoneal tumours, even when lung, ribs and diaphragm are involved [6–8]. Appropriate chest wall and diaphragm reconstruction is necessary after extended radical surgery [9,10].

References