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ASKIN TUMOR: A CASE REPORT AND REVIEW OF LITERATURE

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ABSTRACT

A seven-year old boy with a chronic chest pain, diagnosed with a non-metastatic Askin tumour of the left chest wall. Loco regional assessment in the CT showed a massively necrotic left hemithorax tumour measuring 15cm x 12cm x 13cm. He was treated according to the Euro-Ewing protocol 99 and received an induction chemotherapy then surgery. Given the positive margins, radiotherapy was indicated and delivered in split course using an intensity modulated radiation therapy technique (IMRT) with VAI consolidation chemotherapy. A recurrence of the disease was observed 7 months after the end of the radiotherapy.

KEYWORDS: Askin tumour-Chest Wall-Radiotherapy-IMRT.

INTRODUCTION

Askin tumours is a rare malignant neuroectodermal tumour of the thoraco-pulmonary region with aggressive behaviour and it belongs to the Ewing sarcoma family (ESFT)[1] ESFT are characterized by a re-arrangement involving chromosome 22, and 11. 22 translocations is detectable in more than 95% of cases.[1,2] The prognosis of Askin tumours is very poor with a two-year survival rate of 38%.[3]

Surgery, chemotherapy and radiotherapy or a combination appears to constitute an effective treatment strategy for Askin tumours.

We report a case of Askin tumour with a review of literature.

The Case

A.C is a seven-year old boy, resulting from a non-consanguineous marriage, from a mother of 41 years old and a father of 47 years old. He is a 5th of a sibling of 6. He had no pathological history.

The symptomatology goes back to one year before his consultation in our department. He consulted a paediatrician for left basal chest pain not responding to symptomatic treatment.

A CT scan of the chest was performed and showed a massively necrotic left lung tumour measuring 15cm x 12cm x 13cm with no lymph node metastases (figure 1).

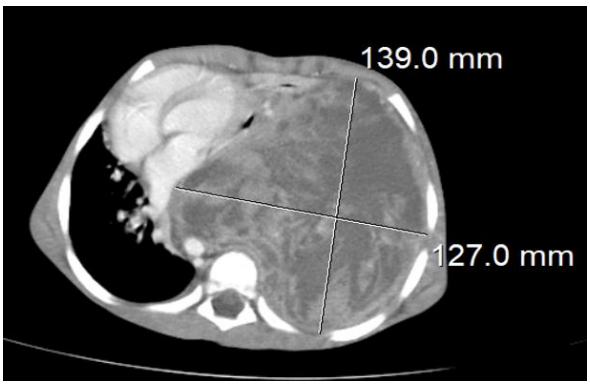

Figure 1: **Axial slice injected thoracic CT of our patient crossing the longest axis of the tumour.**

The pathological examination of the CT-guided biopsy showed a tumour proliferation made of small round cells, with rounded or oval nuclei, fine chromatin and basophilic cytoplasm: PNET EWING group tumour. Complement Fish (fluorescence in situ hybridization) confirmed the diagnosis with rearrangement of the EWSR1 gene.

Thoraco-Abdomino-Pelvic Computed Tomography (CT-TAP) and a bone scan was done which did not show secondary localization.

The patient was treated according to the Euro-Ewing protocol 99: he received an induction chemotherapy with 6 cycles of VIDE (*VINCRISTINE 1.5 mg/m2/day; IFOSFAMIDE 3.0 g/m2/day; plus MESNA;*

DOXORUBICIN 20 mg/m2/day; ETOPOSIDE 150 mg/m2/day).

An evaluation with a thoraco-abdomino-pelvic CT scan was performed after the 6th cycle, and which showed a stability of the tumour.

After multidisciplinary discussion, the patient underwent surgery. On exploration a parietal tumour was found invading the left lung and encompassing the thoracic aorta and the left pulmonary vessels. Total resection of the tumour was performed as well as the adjacent rib with costo-vertebral disarticulation.

The pathological examination and immunohistochemical supplement of the surgical specimen showed a small round cell tumour residue (PNET/Ewing group tumour according to the patient's history). The therapeutic response was estimated at 35%. The resection limits were tumoral and the bone (rib) is not infiltrated by the tumour proliferation described above.

Therefore, in the light of the anatomopathological results of the surgical specimen (R1 resection), radiotherapy with consolidation chemotherapy IVA (*Ifosfamide 3000 mg/m^2/day*, *Vincristine 1.5 mg/m^2*, *Actinomycin 1.5 mg/m^2/day*) (without Actinomycin during radiotherapy) was indicated after the MDT board.

The clinical examination found the patient in a good general condition, good psychomotor development, and a clean scar in the left basi-thoracic region

After two cycles of IVA; an evaluation CT scan was performed (figure 2) showed a tumour residue of about 5 cm in diameter. Radiotherapy was delivered using an intensity modulated radiation therapy technique (IMRT).

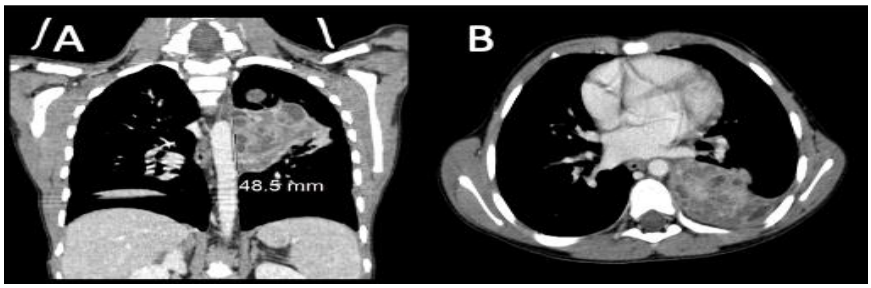

Figure 2: **Injected thoracic CT scan in coronal (A) and axial (B) section of our patient after two cycles of IVA objectifying the postoperative residue of about 5 cm of great axis.**

The GTV was the residual mass after surgery and a 2 cm safety margin as a CTV corrected to anatomical barriers. Areas of scars after biopsy and tumour resection was included. The PTV was generated by a 0,5 cm expansion of the CTV. The volume was reduced to the GTV plus a 0.5 cm margin at 39,6 Gy with a total dose of 55,8 Gy at daily fraction of 1.8 Gy.

There were no significant side effects observed during or in the end of radiotherapy except a moderate skin reaction on the irradiated region.

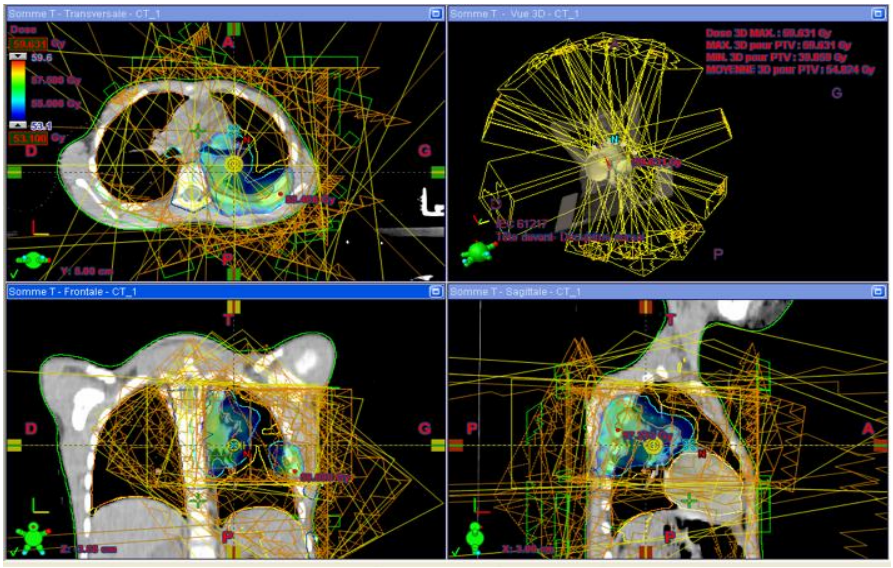

Figure 3: **Disposition of multiple IMRT beams.**

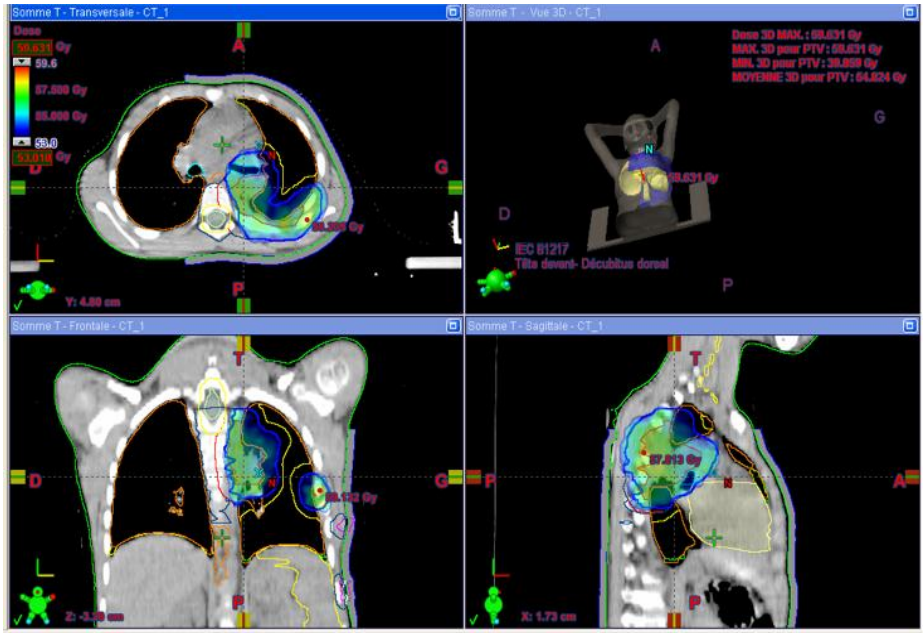

Figure 4: **dosimetry images in the 3 planes of the sum plane showing the 95% isodose conforming to PTV_2 (cyan) which corresponds to PTV boost.**

Seven months following the end of radiotherapy and five months following the end of consolidation chemotherapy, he presented a recurrence with a carcinomatous lymphangitis with a left upper lobar tumour.

After two years follow up, A.C is still alive. He is under palliation chemotherapy (Metronomic protocol) with clinical and radiological stability.

DISCUSSION

Askin tumour is a rare tumour with aggressive behaviour. It was first reported in 1979 by Askin and Rosai.[4] 80% of diagnosed patients are under 20 years of age; however, there are reported cases in elderly patients.[5]

Clinically, the patients who often consult for chest pain associated with a dry cough and dyspnea, evolving in a context of a decline of the general condition with subsequent appearance of an isolated or multiple hard parietal mass.[6]

CT scan or magnetic resonance imaging (MRI) are crucial to determine the extent, the possible pulmonary invasion of the tumour, and invasion of local and distal structures, as well as response to treatment.[2]

Several studies of histogenesis were published. A neuroectodermal origin of this neoplasm was suggested initially, based on some morphologic and ultrastructural findings, but no immunohistochemical analysis was done at that time.[7]

Non-Hodgkin's lymphoma, small cell osteosarcoma, and metastatic neuroblastoma during the histologic examination should be considered differentials.[7]

In immunohistochemistry, there is positivity of both CD99 and Fli-1 and an absence of myogenin expression, lymphoid and neuroendocrine markers. In addition, the cytoplasm is rich in glycogen with dense nuclei.[8]

Askin tumour treatment must include radical resection, radiotherapy, and aggressive chemotherapy.[1] The induction chemotherapy facilitates radical surgery with R0 resection limits.[9] The main role of RT is to achieve a satisfactory control of the primary disease; and it is also administered as adjuvant therapy prior to or following resection. Pre-operative radiotherapy is particularly suitable for patients with a poor clinical response to the initial chemotherapy.[10] It was the case for our patient with poor response after induction chemotherapy and R1 resection.

This tumour is characterized by its local recurrence and metastatic tendency despite the combination of several therapies. The prognosis is very unfavourable, with a two-year survival of 38% and a six-year survival of 14%.[11]

Consent

The examination of the patient was conducted according to the Declaration of Helsinki principles.

Conflicts of Interest

The authors do not declare any conflict of interest

CONCLUSION

Askin tumour is a rare tumour characterized by its aggressiveness and unfavourable prognosis. The treatment is based on a multimodal management, including initial chemotherapy allowing a large surgical resection with negative margins, followed by consolidation chemotherapy. Radiotherapy contributes to the improvement of survival and local control of these patients.

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