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Tumoral Superior Vena Cava Syndrome: About 19 Cases

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Abstract **Original Research Article**

Introduction: Superior vena cava syndrome (SCS) results from obstruction of the superior vena cava and/or its brachiocephalic collaterals by extrinsic compression and/or by tumor or cruoric thrombosis. The aim of this work is to draw up the clinical, radiological and therapeutic profile of SCV linked to cancers whatever their histological types. ***Materials and methods:*** This is a retrospective study carried out at the Military Hospital Mohamed V -Rabat-, over a period of seven years, from January 1, 2011 to December 31, 2017. We included all the patients with histologically confirmed cancers, complicated by SCS. ***Results:*** 16 men and 3 women with cancer presented with SCS during the study period. Their average age was 53 years old. The SCS was indicative of neoplastic pathology in 63% of cases. The most common etiology was lung cancer (58%). The majority of patients received symptomatic treatment (oxygen therapy and corticosteroid therapy). Anticoagulant treatment at curative doses was prescribed in seven patients with thrombosis of the upper case vein and/or its collaterals. None of our patients received a stent or thrombolysis in the event of thrombus. Etiological treatment combined chemotherapy (74%) and/or mediostinopulmonary decompressive radiotherapy (37%). The decrease in SCS was noted in 16 patients (84%). ***Conclusion:*** It is usually a sign of advanced neoplastic disease. Its management, diagnostic and therapeutic, must be as fast as possible and adapted to the cause of the SCS, to the stage and to the histological type of cancer, in order to improve the quality of life of these patients whose prognosis is generally poor.

Keywords: Superior vena cava syndrome, cancer, management.

INTRODUCTION

Superior vena cava syndrome is the clinical expression of the interruption of the superior vena cava in relation to compression, invasion or thrombosis of the vessels. SCV responds to many etiologies which are dominated by neoplastic diseases [1].

The diagnosis is clinical. The severity of the clinical picture depends essentially on the speed of installation of the SCS, which can be the cause of a laryngeal or cerebral edema involving the vital prognosis [2, 3].

Apart from these two situations which require immediate treatment, a biopsy is necessary before the initiation of any treatment. This must be multidisciplinary, integrating radiotherapy, chemotherapy and endovascular treatment, and will depend essentially on the severity of symptoms, type and stage, but also on the general condition of the patient [1].

The aim of this work is to draw up the clinical, radiological and evolutionary profile as well as the methods of management of SCS of neoplastic origin, through a retrospective study carried out at the Military Hospital Mohamed V -Rabat-.

MATERIALS AND METHODS

This is a retrospective study carried out at the Military Hospital Mohamed V -Rabat-, over a period of seven years, from January 1, 2011 to December 31, 2017. We included all the patients with histologically confirmed cancers, complicated by SCS. We excluded patients with a benign SCS.

Data was collected from patient medical records. These data were recorded in a previously established operating sheet. It made it possible to

identify the various epidemiological, clinical, pathological and therapeutic characteristics.

All these data were coded and entered in Excel (Microsoft Office 2016). Data analysis is carried out by the SPSS software. A descriptive analysis of the sample is made; the results are presented in the form of a percentage and an average ± standard deviation.

RESULTS

The group of patients studied includes 16 men (84%) and 3 women (16%), aged 17 to 77 years with an average age of 53 years. Notion of tabagism was present in 13 patients (68%).

SCS was revealing in 12 cases (63%), and occurred in known neoplastic disease in 7 cases (27%). The most frequent clinical manifestations were edema of the upper body, present in 19 cases (100%). Turgor of the jugular veins was noted in 15 cases (79%). Dyspnea in 13 cases (68.4%). thoracic collateral venous circulation in 12 cases (63%) and facial plethora in 8 cases (42%).

The Yale University classification allowed us to classify the severity of SCS [4]. In our series, we found that 43% of cases are classified as grade 1 and 27% of cases are grade 2. For the more severe stages 3 and 4, we noted 15% each. Note that the 3 grade 4 cases progressed to death (Tab. 1).

Table-1: Distribution of patients according to the Yale classification

Yale grading	Number	percentage
Grade 0	0	0%
Grade 1	8	43%
Grade 2	5	27%
Grade 3	3	15%
Grade 4	3	15%

All the patients underwent a thoracic computed tomography scan (CT scan), which the diagnosis of obstruction of the superior vena cava system in all of our patients, therefore a sensitivity of 100%. The CT scan showed thrombosis in 5 cases, extrinsic compression in 12 cases and compression and thrombosis in 2 cases. The CT scan also learned about degree of occlusion and to classify it according to the Stanford's classification [5] (Tab. 2).

Table-2: Distribution of patients according to the Stanford's classification

Stanford's Classification	Number	percentage
Stage 1	8	42%
Stage 2	7	36%
Stage 3	2	11%
Stage 4	2	11%

Lung cancer was found in 11 cases (57%); non-small cell lung cancer in 6 cases (31%) and small cell lung cancer in 5 cases (26%). The other etiologies were represented by lymphoma (2 cases), breast cancer (2 cases), acute lymphoid leukemia (1 case), desmoid tumor (1 case), stomach cancer (1 case) and pancreatic cancer (1 case).

Our patients received symptomatic treatment (oxygen therapy, corticosteroids) and specific treatment (chemotherapy and radiotherapy). The patients in our series did not benefit from the placement of endovascular prosthesis (Tab. 3).

Table-3: Distribution of patients according to the treatments received

Treatment	Number	percentage
Oxygen therapy	17	89%
Corticosteroids	15	79%
Chemotherapy	14	74%
Radiotherapy	7	37%
Endoprosthesis	0	0%
Anti-coagulants	7	37%

In our series, Evolution was favorable in 16 cases (84%). Death occurred in 3 cases (16%).

DISCUSSION

Intra-thoracic cancers are the most common cause of SCS [6, 7]. Malignant SCS are growing rapidly; it takes on average 2 to 3 weeks between its onset and its detection. In this series, SCS was indicative of neoplastic pathology in 12 patients (63%); it appeared during the course of the cancer in 7 patients (37%).

Physical signs are often suggestive. Edema is the earliest sign, it was found in 60 to 83% depending on the series. Turgor of the jugular veins was found in 27 to 92% of patients depending on the series and in 79% in our series. Cyanosis of the cervicofacial territory is rough at the start and becomes general at an advanced stage, it was found in 31 to 45% of cases depending on the series and in 42% in our series. Collateral venous circulation was found in 67% of cases depending on the series and in 63% in our study [8].

Small cell lung cancer is the histological type that provides the most SCS, with variations of 12 to 43% depending on the series [9]. In this study, small cell lung cancer accounted for 26% of cases. It is most often a disseminated small cell carcinoma. The presence of brain metastases when diagnosing small cell cancer would be more frequent when there is a SCS [10]. The other histological types are especially non-small cell lung cancer, notably squamous cell carcinomas which represent 23 to 42%, large cell lung carcinomas which represent 8 to 18%, and undifferentiated carcinomas

which represent 4 to 32% [11]. In our series, non-small cell lung carcinomas accounted for 32%.

Chest radiography has limited value in the positive diagnosis. The thoracic CT scan is required immediately in the presence of a suggestive clinical signs of SCS. Its sensitivity and specificity are close to 100% [12]. In the event of SCS related to lung cancer, CT scan visualizes an invasive mediastino-hilar process, specifies local-regional extension and locates possible secondary pulmonary locations [13]. Magnetic resonance imaging (MRI) is also a powerful technique for the diagnosis of the causal lesion; it differentiates a tumor with vascular invasion from a tumor at the origin of a venous compression with thrombosis on contact but without invasion of the venous wall. Cavography is only used as a second intention after CT and MRI, in the event of a discrepancy between the clinic and these imaging techniques, or when it is the first therapeutic step of interventional imaging [14].

Drug treatment for SCS includes corticosteroid therapy, diuretics, and anticoagulants. Corticosteroid therapy is prescribed for its anti-edematous and anti-inflammatory properties. It is a symptomatic treatment often first-line, administered at doses between 0.5 and 1 mg / kg / day of prednisone; however corticosteroid therapy alone is not effective in the management of SCS [15]. In this series, 79% of patients received corticosteroid therapy from the onset of SCS. The efficacy of diuretics and sodium hydroxide restriction in the treatment of SCS has not yet been demonstrated. The factors predicting the efficacy of anticoagulant treatment in constituted cruoric thrombosis are recent thrombosis associated with an implantable chamber [16]. In the absence of constituted cruoric thrombosis, the usefulness of anticoagulants of the SCS is discussed. Anticoagulants are in this case prescribed in preventive doses. Thrombolysis is recommended if the SCS is related to a recent and occlusive thrombosis of the superior vena cava [17]. In our series, anticoagulants were prescribed systematically in all our patients with superior vena cava thrombosis (37%). Anticoagulant therapy was effective in 5 cases, with clinical improvement after 5-13 days of treatment.

Decompresive radiotherapy has been shown to be an effective treatment for malignant SCS. The success rate varies between 64 and 80% depending on the series. The response is incomplete in about half of the cases and complete in 15 to 23% of the cases [18]. In our series, the results are comparable with those in the literature. One of the treatments for malignant SCS is chemotherapy. The effectiveness of chemotherapy varies depending on the histology of the cancer. This is the treatment of choice when it comes to small cell lung cancer. Response rates vary between series from 62 to 100% and response normally appears within 10 days of chemotherapy administration [19]. In this series, small cell carcinomas responded to chemotherapy in 80% of cases. This response was partial in 4 cases after 3 courses of chemotherapy. Regarding non-small cell lung cancers, as they respond less to chemotherapy, it will not be very effective on the SCS and the response time will be longer [19].

Surgery may be indicated to treat obstructions of the superior vena cava. In case of obstruction related to lung cancer, surgeries can be done for palliative purposes with resection or bypass of the obstructed portion of the superior vena cava without removal of the plmonary cancer, or a curative treatment with resection of the tumor removing the obstructed venous portion. Curative treatment consists of an enlarged pneumonectomy of the superior vena cava with lymph node dissection [20].

The stent is a simple and minimally invasive symptomatic treatment with rapid efficacy on the symptoms. Efficacy is better if the obstruction is due to extrinsic compression than if it is due to tumor thrombosis [21]. In our series, no patient benefited from the placement of a stent.

CONCLUSION

The diagnosis is clinical. The severity of the clinical picture depends essentially on the speed of installation of the SCS, which can be the cause of a laryngeal or cerebral edema involving the vital prognosis. Apart from these two situations which require immediate treatment, a biopsy is necessary before the initiation of any treatment. This must be multidisciplinary, integrating radiotherapy, chemotherapy and endovascular treatment, and will depend essentially on the severity of symptoms, type and stage, but also on the general condition of the patient.

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