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RESEARCH ARTICLE

ELECTRONEUROMYOGRAPHIC AND CLINICAL ASPECTS OF BRACHIAL PLEXUS INVOLVEMENT IN PANCOAST-TOBIAS SYNDROME: A CASE REPORT

Hasna Bentahar, Mohamed Chraa, Najib Kissani and Nissrine Louhab
Neurology Department, Mohammed VI University Hospital, Marrakesh, Morocco.

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Abstract

Pancoast-Tobias syndrome is a rare entity, defined by a clinical triad of cervicobrachial neuralgia in the C8-D1 territory, hand amyotrophy and Claude-Bernard-Horner syndrome, secondary to a tumor of the pulmonary apex. However, there are incomplete or misleading forms that can lead to a wrong diagnosis. The purpose of our observation is to study the clinical and electroneuromyographic patterns of brachial plexus involvement in Pancoast-Tobias syndrome.

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Introduction:-

Pancoast-Tobias syndrome is classically defined by the combination of cervicobrachial neuralgia in the C8-D1 territory, hand amyotrophy and Claude-Bernard-Horner syndrome, secondary to a tumor of the pulmonary apex [1]. It was first described by Henry Pancoast [2, 3]. It's a rare entity that represents less than 5% of bronchopulmonary cancers [2-5]. The clinical symptomatology is the consequence of compression or infiltration of the structures in the vicinity of the pulmonary apex [1]. Therefore neurological manifestations represent one of the circumstances of discovery of TPS, whose different clinical and electrophysiological aspects must be recognized, allowing an early diagnosis for an optimal therapeutic management.

Case Report:-

The patient was a 73 year old male smoker with a 35-pack-year history and no particular medical history. He was referred to our neurology consultation for nine-month history of progressively worsening right shoulder pain radiating down his right arm and into his upper back. The pain was gradually progressive and was associated with weakness and significant weight loss. Neurological examination revealed joint thenar, hypothenar, lumbrical, and interossei muscles atrophy. Muscle strength of the right upper extrimity was 3/5. Motor strength of the left upper extremity and both left lower and right lower extremities was 5/5. Sensation was decreased in the right upper extremity with complete loss at C8–T1 distribution. Examination revealed also right-sided Horner's syndrome with partial ptosis of the right eye, a constricted right pupil and reduced sensation in the right V1 dermatome. Nerve conduction studies demonstrated right brachial plexopathy and sensory polyneuropathy. The sensory nerve conduction studies showed decreased amplitudes in all 4 limbs, probably related to a paraneoplastic sensory neuropathy. F-wave latencies values were in the standards. Electromyography findings showed a neurogenic pattern in the right upper limb muscles. In addition, electromyography comes back normal in the other explored limbs (table 1). Chest x-ray showed right apical pleural thickening (Fig. 1). CT of the chest showed a right apical mass that extended to the right supraclavicular region (Fig. 2). Invasion of the brachial plexus was confirmed by plexus and spinal cord MRI (Fig. 3), while the echodoppler of the upper limb was without anomalies. Percutaneous CT-guided biopsy of the apical mass was performed. Histopathologic diagnosis confirmed non-small cell lung carcinoma.

Corresponding Author:- Hasna Bentahar
Address:- Neurology Department, Mohammed VI University Hospital, Marrakesh, Morocco.

Immunohistochemistry further substantiated the diagnosis of adenocarcinoma. The oncology medicine team was consulted for management of the tumour.

Table 1:- Nerve conduction studies demonstrating right brachial plexopathy and sensory polyneuropathy.

Motor nerve concuction studies					
Nerve/Sites	Lat (ms)	Amp [P] (mV)	Amp [D] (mV)	Vel (m/s)	Onde F (ms)
R Median	**NR**	**NR**	**NR**	**NR**	**NR**
L Median	3.5	5	5	60	-
R Ulnar	2.8	1.1	1	43,5	-
L Ulnar	2.6	8.4	8	63,5	-
R Peroneal	4.2	1.2	1.9	40	-
L Peroneal	3.9	2.9	1.9	37	-
R Tibial	6.4	3.5	3.5		68
L Tibial	6.4	4	4		66

Sensory nerve concuction studies			
Nerve/Sites	Lat (ms)	Amp (mV)	Vel (m/s)
R Median	**NR**	**NR**	**NR**
L Median	1,9	7	64,5
R Ulnar	**NR**	**NR**	**NR**
L Ulnar	**NR**	**NR**	**NR**
R Sural	**NR**	**NR**	**NR**
R Sural	**NR**	**NR**	**NR**

Amp, amplitude; P, proximal; D, distal; Vel, velocity; NR, no response; Lat, latency.

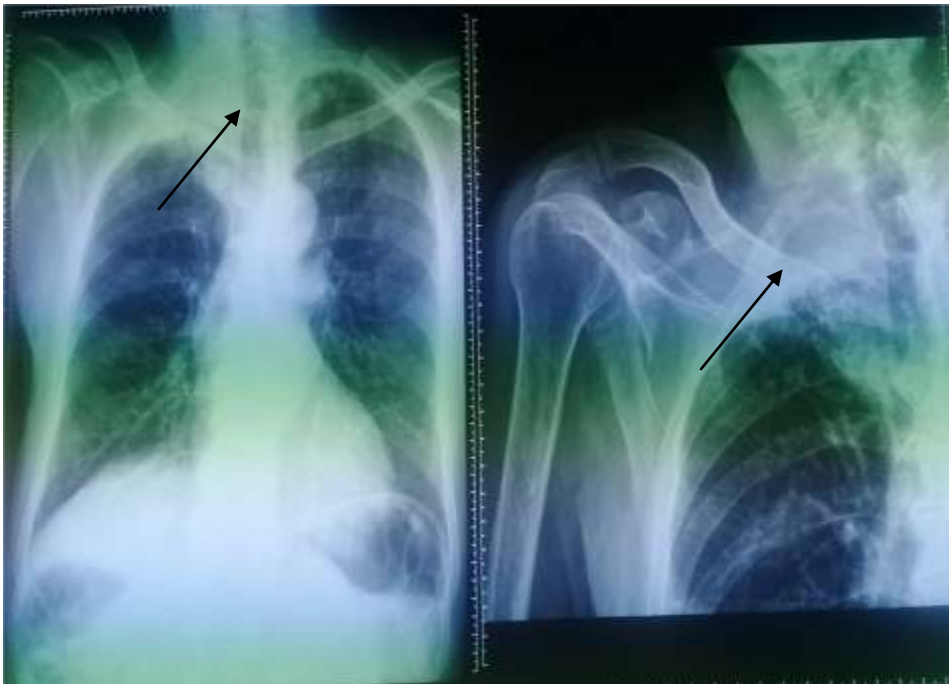

Figure 1:- X-ray of the chest is showing an area of dense opacifcation in the right apical region.

Figure 2:- Axial contrast-enhanced CT scan shows the 6 cm right apical mass.

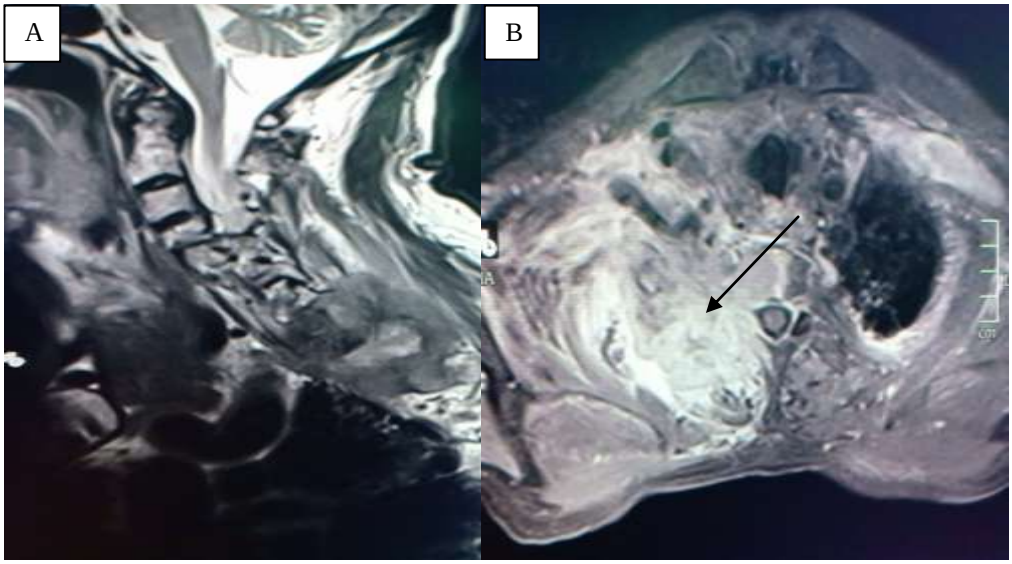

Figure 3:- MRI of the right brachial plexus: sagittal (A) and axial (B) sequences showing diffuse enlargement with marked T2 hyperintensity of the right brachial plexus.

Discussion:-

Superior Sulcus Tumors, frequently denominated as Pancoast tumors may invade any of the structures of the chest apex including brachial plexus, first thoracic rib, sympathetic chain and stellate ganglion near the lung apex [6]. This tumour usually presents with shoulder pain due to invasion of the parietal pleura or brachial plexus [7].

In this observation, the patient's main complaint was pain in the right shoulder, for which an orthopedic or rheumatologic cause would naturally have been suspected first. However, the diagnosis of Pancost-Tobias syndrome was highly likely at the time of the initial consultation, given the history of smoking, significant weight loss, and neurophysiologic evidence of brachial plexopathy. Persistent shoulder pain should always be investigated especially in the absence of local trauma or clinical atypia in smoking patients, in order not to ignore a Pancost-Tobias syndrome [8].

Horner's syndrome is the consequence of an interruption to the cervical sympathetic innervation of the eye [9, 10]. It is a clinical entity specific to brachial plexopathy of neoplastic etiology [11]. In addition to pancoast tobias syndrome, horner's syndrome was also described in association with diffuse large B cell lymphoma, [12] nonHodgkin's lymphoma [13] and extrapulmonary lymphoid granulomatosis [14], the majority of these cases were due to the presence of large adenopathy[11].

There are many neoplastic causes of Pancoast-Tobias syndrome. These tumors are principally non-small-cell lung cancer, with adenocarcinoma as the most frequent histologic type [15-17]. Pancoast-Ttobias syndrome is rarely associated with small cell carcinoma [17]. It should be noted that a wide variety of other entities can also cause Pancoast-Tobias syndrome, including aspergillosis, tuberculosis and lymphoma. However, benign causes are rarely reported in the literature [18, 19].

Data on the diagnostic utility of electroneuromyography in Pancoast-Tobias syndrome are less exhaustive than imaging, in this instance Magnetic resonance imaging (MRI). MRI is recommended for better characterization of suspected structures of the chest apex invasion, including brachial plexus, subclavian vessels, or suspected spine and neural foramina invasion. It provides a more accurate assessment of the local extent of the tumor and the state of nerve and vessel involvement [20]. A study comparing electroneuromyographic findings in patients with neoplastic or radiation-induced brachial plexopathy showed that the distribution of radicular or nerve damage was of little value in predicting the underlying cause of brachial plexopathy. Myokymic discharges are the most contributory element for the etiologic diagnosis [21]. Pancoast tumor has long been implicated as an etiology of brachial plexopathy. The hypothesis of Pancoast-Tobias syndrome should be envisaged not only in the presence of brachial plexopathy, but also when a C8 or T1 radiculopathy is identified [22]. However, if MRI of the brachial plexus remains the reference diagnostic test in Pancoast-Tobias syndrome, the electroneuromyogram can be useful to refine the differential diagnosis in case of unusual clinical or radiological presentations [23].

The treatment of Pancoast tumors for patients able to undergo surgical resection is radical surgery excersion after induction concurrent chemoradiation. Patients with unresectable Pancoast tumors or distant metastasis can benefit from for radiation therapy as palliative treatment [24, 25].

Conclusion:-

Pancoast-Tobias syndrome is not a monolithic entity on the semiological level. There are incomplete forms. However, pain is the most frequent symptom, which in more than half of the cases is typical of cervico-brachial neuralgia. Recognizing the clinical and electrophysiological patterns of neurological damage in Pancoast-Tobias syndrome allows for early diagnosis. MRI of the brachial plexus remains the reference diagnostic test. In difficult cases, an electroneuromyogram may be useful for the diagnosis of atypical presentations.

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