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Spastic Diplegia Following Mediastinal Neuroblastoma

A Case Report

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Cases with neuroblastoma in the mediastinum and spastic diplegia following mediastinal neuroblastoma are reported rather infrequently. To the best of our knowledge, this case report of spastic diplegia following mediastinal neuroblastoma observed in a child is the first of its kind in Turkey. We feel that we have been fortunate in having this child live until today, a period of about five years since his admission into our hospital and in preventing any recurrences.

Case Report

T. Y. (HCH66/8529) - A six month old Caucasian boy was admitted into Hacettepe Children's Hospital on February 22, 1966, with complaints of weakness, fever and coughing as well as profuse perspiration. These symptoms had first been observed three months earlier. One month prior to his admission the patient complained of recurrent fever and dyspnea which were in turn interpreted as bronchitis by his physician. According to his mother, the boy had suffered from dyspnea as early as the first days of life. He was the second child of seemingly healthy parents.

Physical examination : Physical examination on admission revealed moderate pallor. The height and the weight of the patient were above the fifty percentile, 69 cm. and 7.650 kg. respectively. His temperature was 37.5°C, pulse 130 and respiration 28 per minute. His general condition appeared to be fair. There were micro-lymphadenopathy around the cervical and axillary regions. On auscultation breath sounds were normal.

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The day following his admission, the patient ran a temperature of 39.5°C. A throat culture taken on that day revealed Beta-hemolytic streptococcus.

Laboratory Findings : The chest X-rays revealed a large round mass with possible fine calcifications in the superior posterior mediastinum. (See Figure 1) The X-rays of the skull, long bones, vertebrae and kidneys seemed to be normal.

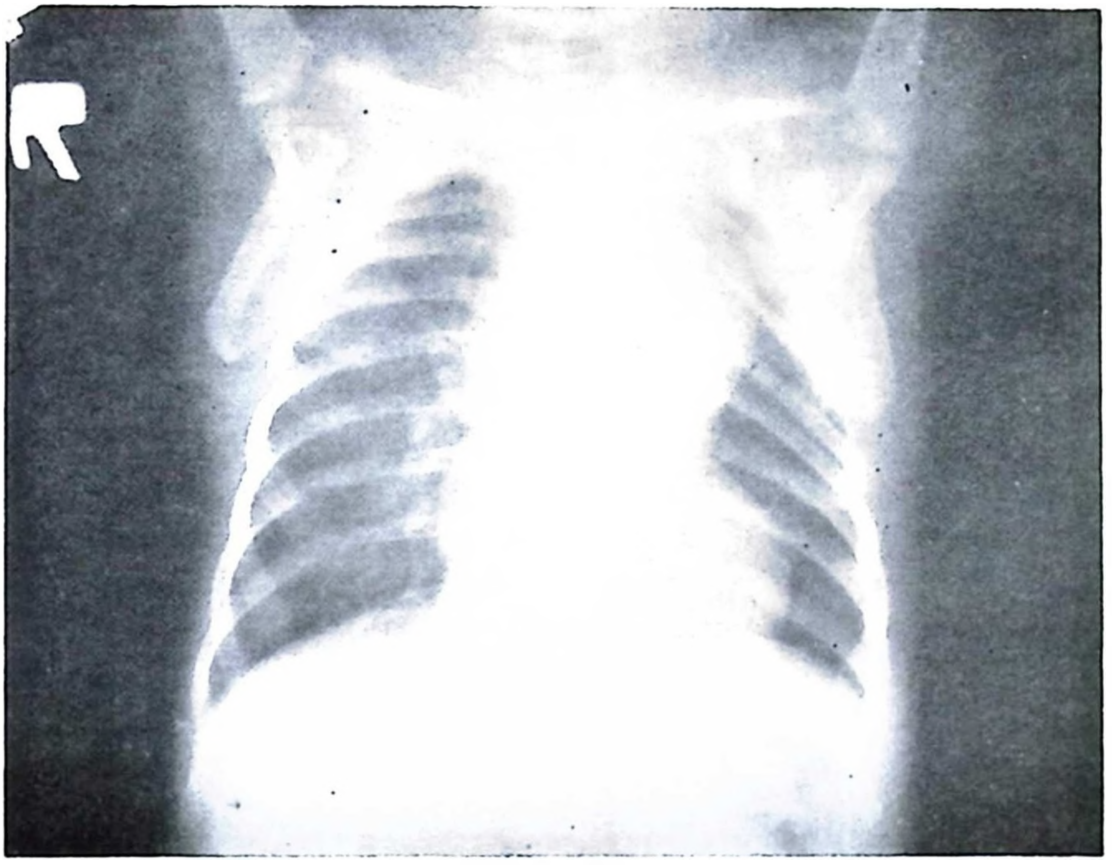

Figure 1

Hemoglobin was found to be 8.60 gr/100 ml, WBC 6,200 per cu. mm. with 48 per cent neutrophils and 52 per cent lymphocytes. Platelets were observed to be numerous in peripheral smears. ESR was 43 mm per hour. The bone marrow was normocellular with a normal M/E ratio and rosette formations were not seen. VMA (3-methoxy-4-hydroxymandelic acid) was found to be 5.6 microgram/mgr creatinin (normal 0-6 microgram). The biopsy taken from the tumor mass revealed neuroblastoma. The lymph node was infiltrated with neuroblastoma cells. The biopsy of the rib bone revealed no metastasis.

Clinical course : The patient was hospitalized for twenty days and was initially given a course of penicillin and streptomycin for treatment of the Beta-hemolytic streptococci in his throat.

Under general anesthesia thoracotomy was performed on the left side. The tumor mass extended from the second to the sixth or seventh rib, medially from the vital mediastinal organs such as the aorta, laterally to the middle axillary line. It seemingly infiltrated the chest wall, mediastinal lymph nodes as well as other mediastinal structures. It appeared to be hard and calcified and was found to be inoperable. Biopsies were taken from the tumor mass, mediastinal lymph node and the rib bone.

It was then decided to give radiation therapy. The patient was given a total of 1,800 r to the chest. Following this therapy the tumor mass had almost disappeared. On his discharge from the hospital, the patient was prescribed Endoxan (Cytoxan); the drug was discontinued by his parents, however, a few days after the patient's return home.

During his stay at the hospital, the boy developed flask paraplegia in the lower extremities. Shortly after radiation therapy the paralysis became spastic. Control of anal sphincter had never been impaired. At that time, the survey of the bones, including the vertebrae, showed no metastasis. As mentioned above, the biopsy from the rib bone was free of metastasis. Because of the improvement of the patient, tests such as spinal fluid examination or myelography were not performed. This neurological defect was thought to be due to sclerosis following radiation therapy to the possible metastasis in the medulla spinalis. However, this was not proved. Upon discharge from the hospital, he was advised to take physiotherapy and massage treatment in the lower extremities.

The patient comes regularly for control to our out-patient clinic. He started to talk at the age of 1.5 years and was able to sit when about 3 years old. From the date of his discharge until his last control examination on July 13, 1970, chest and bone X-rays revealed no metastasis (see Figures 2 and 3) and his blood count was found to be normal.

The WMA was repeated on July 13, 1970 and was found to be 1.35 migrogram/creatinine.

The patient's latest control examination was carried out on July 13, 1970, when he was five years old. (See Figure 4) He was observed to be a fairly well developed boy with the height and the weight around ten and twenty-five percentiles respectively. He had a deformity on the left side of his chest close to the posterior portions of the third and fourth ribs, with pronounced kyphoscoliosis. He showed a prominence over the anterior portions of the ribs on the right side of his chest. His upper extremities were normal. Both hips could only be

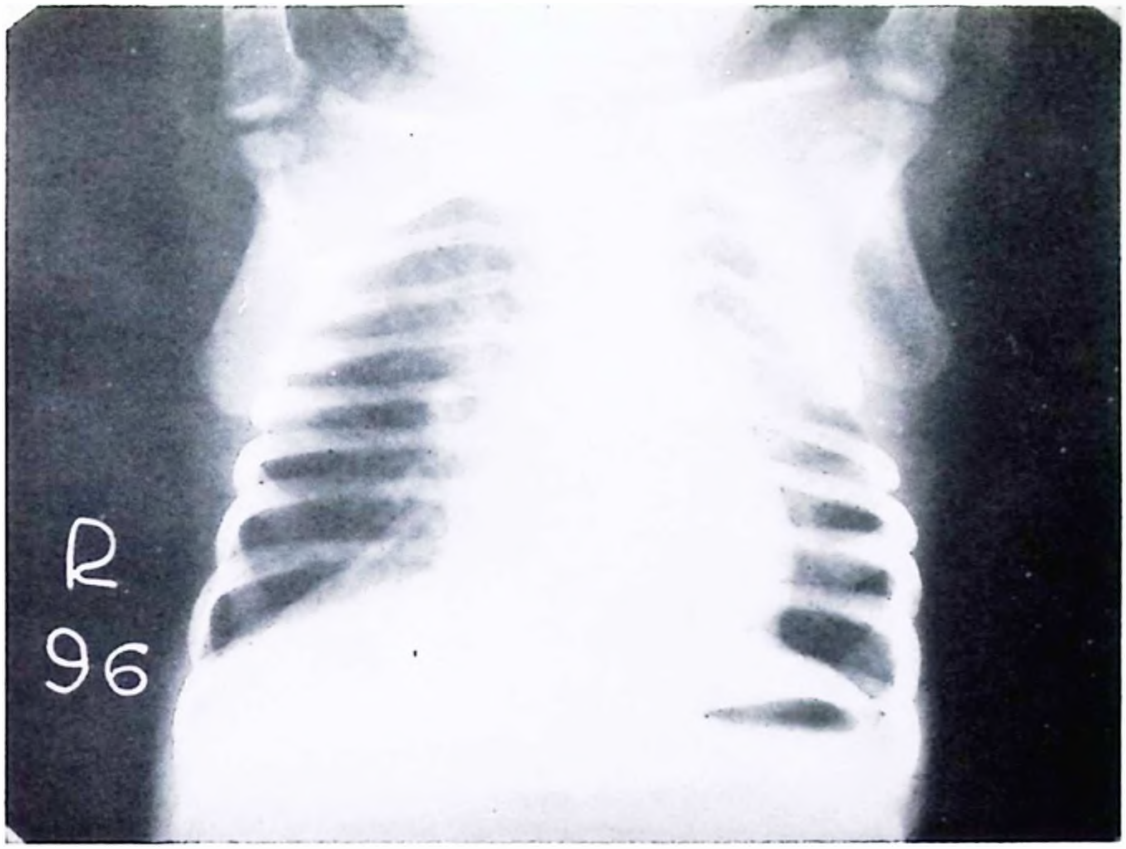

Figure 2

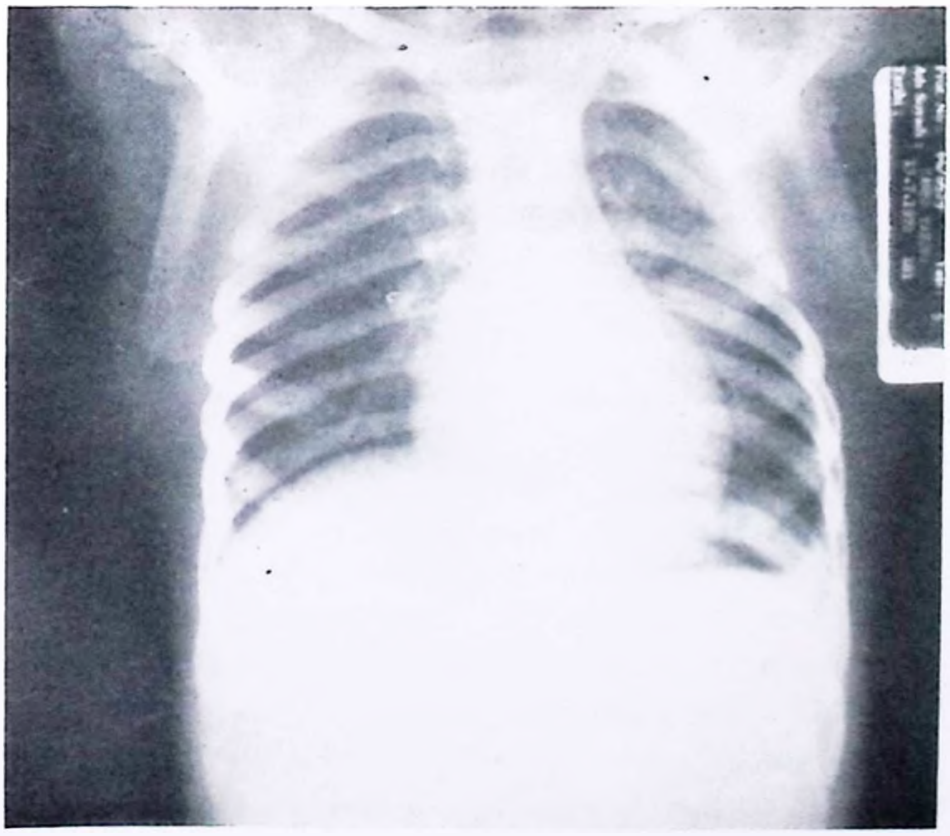

Figure 3

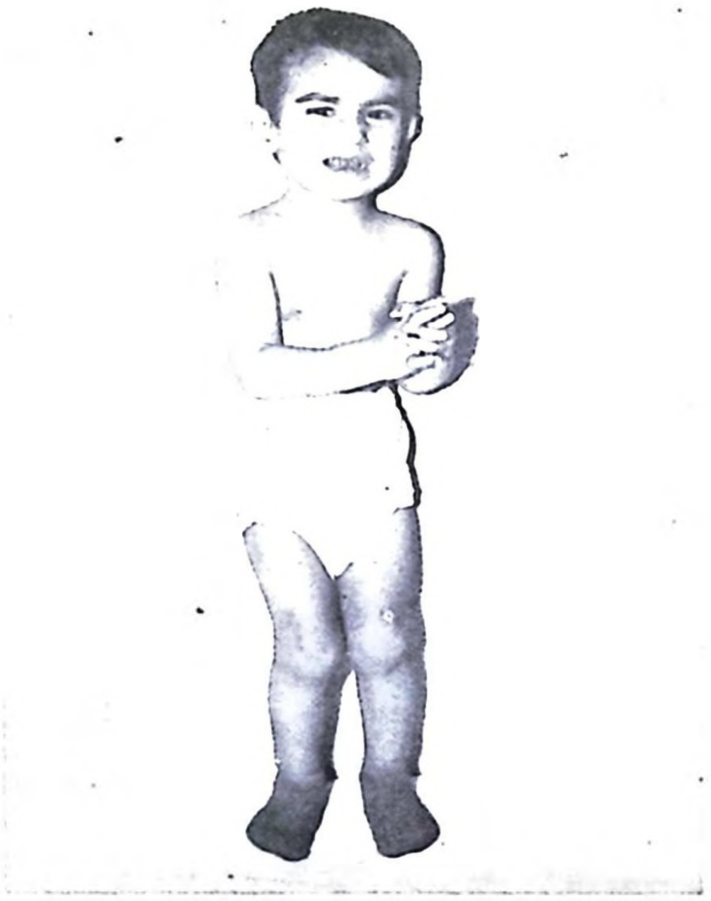

Figure 4

abducted up to 30°. A 20° contracture extension was spotted in both kness. Right and left feet indicated an extension of only 100°. Pes planus were noted in both feet. Deep tendon reflexes were active. Babinsky was positive on both sides and clonus was positive only on the right side. The patient had increased muscle tone. Coordination was found normal. A sensory check-up of the patient was not available. The child was able to walk slowly by sliding his feet and placing one foot in front of the other.

Discussion

Previous to the operation for a tumor in the mediastinum, we had definitely established the patient's ailment to be flask diplegia. Shortly after the operation, while he was receiving radiotherapy, the child developed spastic diplegia. Observing the present healthy condition of our patient and the fact the tumor has never recurred, we have speculated the probabilities for the cause of diplegia to be :

1. Sclerosis around the medulla spinalis secondary to radiation therapy,

2. Occlusion of periaortic vessels as a result of radiotherapy,

3. Infiltration of the tumor in the medulla spinalis. (This possibility was regarded as the least likely).

We believe that a comparison of our case with those found in the literature will be of interest to our colleagues. According to Alpert et al[4] neurological disturbances almost always are secondary to metastasis or invasion of the nervous system. However, primary neuroblastomas involving the central nervous system have been reported. Dargeon[6] has published several reports of cases with neuroblastoma including metastasis of the nervous system.

Cranial nerve palsies, proptosis, increased intracranial pressure and cord compression may be attributed to the metastasis of the tumor present in the nervous system. There have been many reports of spinal cord dysfunction, manifested generally as cord compression secondary to extradural tumor. Spinal cord involvement may occur following invasion of the spinal canal through the intervertebral foramina or by metastases. Cross et al[5] reported that of the 30 cases with cord neoplasm in children, six were extradural neuroblastoma. Alpert et al[4] reported the spinal cord metastases to be common and noted 13 of their neuroblastoma cases they observed as affecting the spinal cord, occuring in the form of extradural deposits which were associated occasionally with metastases of bones.

According to Carey et al[1], weakness, disturbance of gait or incoordination of the lower extemities, or all three, were noted in patients with neurogenic tumors. Besides the above mentioned findings, scoliosis was a prominent feature of these tumors. One of the benign ganglioneuroma cases mentioned was reported to have multiple operations for removal of a tumor which had presumably occurred within the spinal canal and extended into both sides of the thorax. This particular patient had spastic paralysis and contracture deformities of the legs, four years after the operation. None of the neuroblastoma cases reported by these authors developed spastic paralysis of the legs following radiation therapy.

Katzman et al[2] points out that the more severe skeletal changes seem to occur in patients of two years of age or younger at the time of the original radiation treatment and when the dosage exceeds 2,000 r. The observance of the relationship between the age of the patient and the dosage of radiotherapy may make a much better prognosis possible

for patients under two years of age treated for neuroblastoma. As observed in our cases, scoliosis was one of the most common sequelae encountered in cases reported by these authors following radiotherapy.

Anderson[3] noted a case of thoracolumbar neuroblastoma with complete paraplegia who had metastasis in the spinal cord. The tumor in this case was excised subtotally and one course of radiotherapy (3,000 r.) was given which resulted in the steadily lessening disability. Six and a half years after the treatment no impairment was observed in the function of the legs.

Eklöf[9] reports that roentgenologic evaluation of patients with primary intrathoracic neuroblastoma, indicate calcification to be uncommon in such cases. Calcification is not seen at all in cases of secondary mediastinal deposits and, most important, what is clinically presented as a primary intrathoracic mass, may in reality, represent a metastatic focus. Our case on the other hand, shows a certain amount of calcification in the biopsy which could point to a nueroblastoma with primary origin. Eklöf also asserts that primary intrathoracic cases produce earlier symptoms than abdominal cases.

Summary

The clinical course of a case with mediastinal neuroblastoma including spastic diplegia which afflicted a six-month old boy has been presented and pertinent material in the medical literature has been discussed in an effort to compare our findings with those of our colleagues and to emphasize the significance and the necessity of early treatment in similar cases.

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