Case Report

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Abdominal expansion using a polytetrafluoroethylene prosthesis in the treatment of Pepper syndrome

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Abstract The authors report three cases of stage IV-S neuroblastoma in infants aged 4, 6, and 8 weeks, who despite chemotherapeutic and radiotherapy required surgical intervention to urgently relieve major thoracoabdominal compression secondary to massive hepatomegaly. The results were successful, with abdominal expansion being achieved by the introduction of a polytetrafluoroethylene prosthesis, which was removed during the 2nd, 3rd, and 7th postoperative months, respectively, after tumor regression. Two children were in complete remission 32 and 38 months later, the 3rd died after 16 months of tumor progression.

Key words Neuroblastoma · Stage IV-S · Hepatomegaly · Abdominal decompression

Introduction

Stage IV-S neuroblastomas (NB), or Pepper syndrome, are tumors that despite metastatic diffusion have a good prognosis before the age of 1 year due to the possibility of regression and spontaneous tumor maturation. Mortality is as much related to mechanical complications due to hepatomegaly as it is with tumor progression. We report three cases of abdominal expansion with the aid of a polytetrafluoroethylene (PTFE) prosthesis in infants presenting with Pepper syndrome and whose survival was in jeopardy as a result of major thoracoabdominal compression.

Materials and methods

Three female infants presented with Pepper syndrome at 4, 6, and 8 weeks of age, respectively. Computed tomography revealed a metastatic liver tumor that occupied the entire abdomen (Fig. 1) and, in two cases, a primitive adrenal tumor. The lesions showed hyperstaining on MIBG (methyl-iodo-benzyl-guanidine) scintigraphy. There was no medullary invasion. Blood and urine examinations revealed elevated levels of catecholamines in all three cases, confirming the secretory nature of the NBs.

In the 1st infant, because of tumor progression and the appearance of signs of pulmonary compression, treatment combining chemotherapy (vincristine and cyclophosphamide) and hepatic radiotherapy (three sessions of 1.5 Gy) was investigated. Before hepatic irradiation was completed, the compression worsened with signs of inferior vena cava (oliguria, edema of the lower limbs) and chest compression, which necessitated assisted ventilation. After 4 days, after the development of cardiac and renal failure, emergency surgical decompression was carried out comprising abdominal expansion using a PTFE plate. The intervention allowed immediate decompression of the thorax and abdomen, improving the hemodynamic and ventilatory parameters by reducing the pressure on the vena cava and pulmonary re-expansion. Tumor reduction began on day 10 and extubation was possible on day 22. The PTFE plate was removed during the 7th month because of partial cutaneous dehiscence. There were few adhesions, and the residual hernia was treated using a resorbable polyglactin prosthesis. At 34 months a right adrenalectomy was performed. At 38 months this child was considered to be in complete remission, the only consequence being a simple deformation of the thorax with projection of the costal hood.

In the 2nd infant, chemotherapy followed by the same protocol was instigated 4 days after diagnosis because of rapid progression of the tumor. On the 9th day,

Fig. 1 Abdominal CT scan showing massive hepatomegaly.
the appearance of a compressive syndrome associated with a pulmonary infection by vancomycin-resistant staphylococci necessitated transfer to the intensive care unit. On the 15th day the clinical picture deteriorated, and abdominal expansion was performed followed by immediate postoperative hepatic radiotherapy. Tumor regression allowed extubation on day 15. Removal of the PTFE plate was performed on day 40 in the absence of any continuing compressive signs because of partial cutaneous dehiscence. The abdominal wall was repaired using the same technique as in the first case. Follow-up was marked by a relapse at 7 months with bone metastases (leading to Hutchinson’s syndrome, which was the cause of death at 16 months).

In the 3rd case, chemotherapy was begun because of rapid tumor progression. Five days after the start of treatment, abdominal expansion was performed followed by hepatic irradiation (3 sessions of 1.5 Gy on days 1, 2, and 3) because of severe clinical deterioration. Due to continuing tumor progression, a second series of hepatic irradiations was performed (3 sessions of 1.5 Gy on days 15, 16, and 17) along with 2nd-line chemotherapy associating Vp 16 and carboplatin. The tumor regressed, which allowed extubation and removal of the plate on day 90. Repair of the abdominal wall was performed in the same way as in the first two cases. After 32 months’ follow-up, the child was in complete remission.

Discussion

Pepper syndrome is a particular clinical form of metastatic NB of the newborn. Described for the first time by Pepper in 1901, the clinical picture is dominated by marked abdominal distention secondary to massive hepatomegaly [11]. In Evans’ classification of neuroblastomas [5], it is regarded as stage IV-S with mainly hepatic involvement (stage IV-S = localized, primitive stage I or II tumor [not exceeding the median line] in patients younger than 1 year with metastases limited to the liver, skin, or bone marrow with no radiologically detectable bone lesions) [11, 13]. It represents the most common form of neonatal NB (30% during the 1st month of life) and affects boys and girls equally [8]. Before the age of 1 year, despite metastatic spread, because of the possibility of regression or spontaneous maturation these tumors have such a good prognosis that survival at 5 years is approximately 80%.

Death is as much related to mechanical complications as to tumor progression [14]. In fact, the compression caused by the hepatic volume may cause respiratory failure due to upward displacement of the diaphragm and pulmonary compression, renal problems due to compression of the inferior vena cava, and cardiac problems due to reduced venous return.

The theoretical therapeutic approach is the least aggressive possible in infants less than 1 year of age, and consists of close monitoring (clinical, biological, and radiologic) in the absence of tumor progression in the hope of spontaneous regression. However, the picture can deteriorate rapidly; the hepatic volume and resulting compressive phenomena can become life-threatening, necessitating fast, aggressive action. Hepatic radiotherapy at the recommended schedule of 3 sessions of 1.5 Gy is considered the treatment of choice with the objective of reducing hepatic volume rather than controlling tumor mass [13, 15]. Chemotherapy (cyclophosphamide and vincristine) is used for diffuse tumor growth, more or less combined with hepatic radiotherapy. However, the response to this medical treatment is uneven and may be slow [2]. For this reason, surgical treatment involving abdominal decompression may be necessary, sometimes as an emergency, in cases where the condition has become life-threatening as a result of compressive phenomena before the tumor has begun to regress.

Since 1975, Schnaufer and Koop, have used Silastic prostheses inserted into the abdominal wall to create a ventral abdominal hernia [12], a technique identical to that using a silo described by Allen and Wrenn for the treatment of gastroschisis [1]. This surgical treatment is controversial. The theoretical advantage consists of immediate decompression by increasing the volume of the abdominal cavity, but without any effect on the tumor volume. The problem with this type of surgery, which involves creating a hernia covered by an external Silastic patch, is that morbidity and mortality are very high, most often as a result of infection in an immunosuppressed patient. A review of the literature shows that among 11 patients subjected to this intervention under the same clinical conditions, there were 9 postoperative deaths: only 1 was from tumor progression, the others were due to infection. This technique was reserved for only the most desperate cases [2–4, 6, 7, 10, 12].

In the three cases reported here, we performed abdominal expansion with a PTFE plate in accordance with a technique described by Lee and Applebaum in 1994 [9]. The intervention begins with a transverse abdominal incision. Very wide subcutaneous detachment exposes the musculo-aponeurotic surface, in which a cruciform incision is made horizontally up to the large muscles and vertically from the xiphoid to the pubis, which brings about immediate decompression of the abdomen with protrusion of the liver tumor. Repair of the wall is performed with the aid of a PTFE plate, which covers the musculo-aponeurotic defect. This plate is positioned subcutaneously (Fig. 2) after passive stretching of the cutaneous edges to allow a skin suture without tension, with aspirated drainage.

There seem to be many advantages to this technique of abdominal expansion. It is an intervention that can be rapidly and simply performed, and it has an immediate beneficial effect on the compressive symptoms. Compared to the technique using uncovered Silastic

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