The case report of Askin’s tumor in a 16-year-old girl is focused on the still debatable surgical controversy in the treatment of PNET tumor, i.e. whether disarticulation of involved rib at the costovertebral joint should be accepted as the mandatory surgical procedure. It was concluded that the procedure, if feasible, may offer better prognosis of PNET because progression-free survival rate of patients without costovertebral junction involvement reported in multicenter studies was statistically significantly better than in patients in whom PNET has involved the costovertebral junction or bone metastases were present at the diagnosis. The cartilage is a natural barrier for tumor spread and this property should be augmented by radical surgery. Disarticulation of involved rib or ribs and pleurectomy should be routinely performed if the surgery is contemplated with proper timing between the cycles of induction chemotherapy.

Key words: Askin’s tumor, PNET, rib disarticulation, pleurectomy.

INTRODUCTION

Ewing’s Family Tumors (EFT) comprise several infrequent but highly aggressive tumors of primitive neuroectodermal origin, occurring in pediatric age (Primitive Neuroectodermal Tumor - PNET, Askin’s tumor - a PNET of the chest wall and Ewing’s sarcoma). Histologically, the tumors have small round-to-oval cells and a lobulated stroma with some degree of neural differentiation. Due to its infrequent occurrence single institution surgical experience is limited to reports of several cases and the surgical treatment of choice is not known since no clinical series with with surgical therapy have been reported. Issues regarding diagnosis and therapy have been uniformly accepted. The first multicenter randomized study demonstrated improved event-free survival for chemotherapy and radiotherapy compared with radiotherapy alone. Still, the study did not include surgical aspects. A core needle biopsy is sufficient for histological diagnosis and an open biopsy is required when core biopsy has failed or in cases with superior vena cava syndrome due to very large masses. Fine needle aspiration cytology is not recommended for diagnosis as the amount of cells obtained may not be adequate and should be reserved for confirming metastasis or recurrence of tumor. Regarding surgery, there is still a controversy whether rib disarticulation is needed in all children with PNET. Due to various clinical presentation at the time of diagnosis and the wide range of tumor involvement of thoracic wall tissues (muscles, ribs, pleura, lungs), as well as insufficient single institution experience, it is still a matter of debate if rib cartilage can act as a natural barrier to tumor spread. The properties of cartilage; its lymphatic and vascular supply are well recognized as a barrier to the spread of i.e. both infection and tumors. The fact is supported by event-free survival studies in PNET; a 5-year survival rate of patients without costovertebral junction involvement was 66%, whereas patients with PNETs involving the costovertebral junction had a 21% 3-year survival. The difference was statistically significant (p = 0.01). The 5-year progression-free survival rate of patients without costovertebral junction involvement was 58%, whereas patients with PNETs involving the costovertebral junction had a 14% 1-year progression-free survival (p = 0.004). The study clearly supports the assumption that costovertebral junction is the ultimate barrier preventing further spread of malignant cells alongside the affected rib. The term “intralesional excision” is used when only partial resection was feasible, the prognosis being very poor. On the other hand, it has been proposed that the lesion should be completely removed, but extensive resections or rib disarticulation are considered not necessary and should be administered when resection margins are microscopically
positive and the tumor shows definitive invasion of costovertebral junction.

**CASE REPORT**

A 16-year-old girl was referred with large mass in the right thoracic cavity. It was obvious that the tumor must have grown for months with minor symptoms (episodes of cough attributed to respiratory infections). Unfortunately, the mass could have been recognized without any sophisticated imaging; on admission there was dullness in the right chest and completely absent breath sound on auscultation. CT scan demonstrated a large mass that literally occupied the whole right chest cavity (Figure 1). The diagnosis of PNET was established with core - needle biopsy under sedation and analgesia. General anesthesia was deliberately avoided having in mind risks associated with extended involvement of mediastinal structures. Induction chemotherapy comprised five cycles of CWS - 96 protocol; High risk - arm B - CEVAIE; Holoxan (ifosfamide), VP-16, Actinomycin D, Oncovin, Carboplatin and Epirubicin.

Repeated CT after the fifth cycle demonstrated excellent response to chemotherapy (Figure 2). It was decided that radical surgery could be performed at the time. Upon surgery, a remaining mass was attached to the fifth rib. Only minor adhesions to the visceral pleura and thickening of the parietal pleura were found. PNET did not invade adjacent ribs or thoracic muscles. The fifth rib was detached from its sternal attachment and disarticulated in costovertebral junction (Figure 3). Thickened parietal pleura was easily detached from the inner thoracic wall and widely resected for histopathology examination. The remaining defect was easily covered with the muscles of the thoracic wall and no prosthetic material was needed to repair the defect. The microscopic examination demonstrated advanced PNET necrosis and scattered microscopically verified focuses of viable malignant cells in the mass and in the parietal pleura. There were no viable PNET cells in the rib and the cartilage was of normal appearance and tumor free, as well as all resected margins. The patient recovered uneventfully and was referred to the hematology/oncology department for further chemotherapy and radiation therapy.

**DISCUSSION AND CONCLUSION**

The case report has been focused on the controversy of radical surgery in pediatric patients with PNET tumor of the chest wall. Although the term Askin’s tumor is still widely used to describe PNET of the chest wall, originally thought to be distinct from Ewing’s, it is now considered a as a member of Ewing’s Sarcoma Family Tumors. Rib disarticulation was done previously in three more patients; Unfortunately, Askin’s tumor was resectable at the admission and the patients are tumor free after 3 years. Another 6 patients from this institution had extensive PNET involvement of the thoracic wall and only partial resection was possible, i.e. “intralesional excision”. Their response to chemotherapy and radiation therapy was poor.
motherapy makes radical surgery possible and markedly decreases tumor vascularity. An experienced surgeon is supposed to evaluate CT between chemotherapy cycles to determine optimal timing for reasonable high probability of radical resection of PNET. Poor response to induction chemotherapy makes surgery only a desperate attempt to contribute to prolonged disease-free survival. Unless the regimen is changed in future it deserves no elaboration at the moment, being uniformly accepted; induction chemotherapy makes surgery only a desperate attempt to contribute to prolonged disease-free survival. Radiotherapy should follow resection of the primary tumor with accurate marking of the vertebral joint, as seen in the reported case. Furthermore, it has been proven that better survival is expected with lung involvement compared with those with bone metastases. Besides, surgical experience with rib resection makes it rather illogical to preserve several centimeters of the resected rib, particularly where the rib is affected very close to the vertebral joint, as seen in the reported case. Furthermore, very high malignancy and poor prognosis in most of the cases of PNET of the chest wall should guide the surgeon to be as radical as possible; the properties of the cartilage as a natural barrier being the additional support to this approach. Retrospective surgical reports from single institutions have already been published and reviewed; further prospective studies are limited by ethical issues: if a researcher believes that rib disarticulation can contribute to higher survival rate, he or she may not create a control group of patients in whom rib disarticulation would be deliberately avoided. Another suggested surgical approach is pleurectomy as wide as possible; it is reasonable to believe that large initial masses were in close contact to parietal pleura and might have left microscopic PNET cells. Therefore, it seems very logical to resect parietal pleura as wide as possible - the procedure being easy (often performed for the treatment of repeated spontaneous pneumothorax before the introduction of less aggressive thoracoscopic pleurectomy). Even minor microscopic evidence of PNET within parietal pleura (as observed in this case) is important for planning radiotherapy.

In conclusion, all relevant data and single institution experience suggest that rib disarticulation should be accepted as a mandatory surgical procedure, as well as wide resection of parietal pleura, far off the primary PNET. Further molecular genetics studies influencing the prognosis of the disease may change radical surgical approach to PNET, the way genetics did it in neuroblastoma.

SUMMARY

DEZARTIKULACIJA REBRA I PLEUREKTOMIJA KOD ASKINOVOG TUMORA: OBAVEZNA PROCEDURA U RADIKALNOJ HIRURGIJI KOD DECE? PRIKAZ SLUČAJA I SMERNICE ZA HIRURŠKO LEČENJE

Prikaz Askinovog tumora kod šesnaestogodišnje devojčice je usmeren ka nerazrešenoj kontroverzi u hirurškom lečenju PNET tumora; da li dezartikulacija rebra u kostovertebralnom zglobu treba da se prihvati kao obavezna hirurška procedura?

Zaključeno je da ovom procudrom, ukoliko je izvodljiva, može da obezbedi bolja prognoza PNET tumora jer je multicentričnom studijom dokazano da postoji statistički značajno bolja prognoza kod bolesnika sa PNET tumorima ako kostovertebralni zgob nije zahvaćen tumorem ili ako ne posjete metastaze u kostima. Hrskavica predstavlja prirodnu barijeru za progresiju tumora i hirurgijom treba da se pospeši ova njena osobina. Dezartikulacija zahvaćenog rebra ili više rebara i pleurektomijsa treba rutinski da se uradi ako se hirurško lečenje planira u pravo vreme izmedju ciklusa indukcione hemoterapije.

Ključne reči: Askinov tumor, PNET, dezartikulacija rebra, pleurektomijsa.

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Acknowledgment:
The authors are deeply grateful to Dr Bruno De Bernardi, the head of Hematology/Oncology department, Istituto Giannina Gaslini, Genoa, Italy and the moderator of SIOP neuroblastoma project, for his invaluable personal communication, his advises and persistence on surgical aspects reported here.