Editorial



# Thymomas in children

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The treatment of thymic epithelial tumors (TETs) in children may pose a serious problem. This is because thymic tumors—thymomas and thymic carcinomas, are extremely rare in patients of this age group, and thus a consistent treatment scheme has not been established. The most widely used European or American guidelines were established for adult population and do not include detailed instructions for paediatric oncologists (1,2). Such instructions may be found in a paper of Sigurdson *et al.*, where the authors presented a multidisciplinary expert discussion regarding the case of an 11-year-old girl with B2 thymoma (3). Both the diagnostic and the therapeutic aspects were considered in context of the young age of the patient.

The international literature encompasses only case reports and a few small series of paediatric cases with TETs (4-10). One of the most numerous series, which included 36 children (from France, Italy, Germany and Poland) with TETs was reported by the European Cooperative Study Group for Pediatric Rare Tumors (4). There were 16 thymomas (mainly B1) and 20 thymic carcinomas in the analyzed cohort. In the thymoma group the authors noted a slight female predominance and a median age of 11 years. Most of the tumors were resected and a complete resection with clear surgical margin (R0) was achieved in almost all cases. In three cases surgery was preceded by neoadjuvant chemotherapy. Two patients died, one from complications of the treatment and another from the progression of the disease. The rest were alive with no evidence of disease after a mean follow up of 5 years. Patients with thymic carcinomas were usually males with a median age of 14 years. In most cases [16] the disease was diagnosed at an advanced, inoperative stage, but surgery was performed on nine patients after initial chemotherapy. The resection was complete (R0) in five cases. Most of the patients were treated with various multidrug regiments and/or received radiotherapy. Fourteen patients died due to the progression of the disease and one due to complications from chemotherapy. The median follow up with no evidence of disease was 2.8 years (4).

Carretto *et al.* presented a series of nine patients with TETs registered within 9 years in Italy for the nationwide project *Tumori Rari in Età Pediatrica* (Rare Tumours in Paediatric Age) (5). The cohort comprised of four males and five females with a median age of 12.4 years. There were five thymic carcinomas and four thymomas (mainly B1) in the group. The clinical course of all the thymic carcinoma cases was unfavourable—the disease progressed and the patients died within one and half year after diagnosis. Three out of four thymomas were in low stages, and they were completely resected. They did not revealed progression or recurrence from 22 to 93 months of observation after diagnosis. The fourth thymoma was detected at a very advanced stage with multiple metastases and the patient died even before the establishing of a final diagnosis (5).

Gun *et al.* in an analysis of mediastinal tumors in children who were operated on in one institution over a 26-year period collected only four cases of TETs: two thymomas (subtypes unknown) and two thymic carcinomas (7). Liu *et al.* in a group of 409 mediastinal lesions did not find TETs in the paediatric population at all (11).

In context of these papers the patient presented by Sigurdson *at al.* showed similar epidemiological profile as other young thymoma patients—a female at the beginning

of her second decade of life (3). The histological subtype differed slightly (B2) but the difference may be the result of modifications to the histological criteria of the current thymic tumors classification, that limited the number of diagnosed B1 tumors. These cases are presently more often classified as B2 thymomas (12).

Thymomas are in adults the most numerous subgroup of TETs, and thymic carcinomas constitute about 20% of these tumors (12). In the studies presented above, based on the paediatric population the contribution of thymic carcinomas in all TETs was clearly higher (about 50% or more) (4,5). Thymomas, both in children and adults, usually have a much better outcome than thymic carcinomas, provided that the tumors are in a low stage, and that they are completely resected (R0) (4-6,12).

After surgery, depending on the invasiveness of the neoplasm and the completeness of the resection, adjuvant radiotherapy may be considered (1,2). For the 11-year-old patient described by Sigurdson *et al.* the experts did not propose adjuvant radiotherapy. They argued that the potential risk of adverse effects of radiation in paediatric patient exceeded the potential oncological benefit (3). The patient underwent surgery twice, but eventually a complete resection (R0) was achieved. The tumor had invaded the adipose tissue, but the extend of the invasiveness was low (stage IIA in Masaoka-Koga staging system and pT1a/I in pTNM classification). Iorio *et al.* described a similar case of a 7-year-old boy with myasthenia gravis and B3 thymoma in Masaoka stage II. The tumor was completely resected, and after surgery the authors decided not to perform adjuvant radiotherapy for the same reasons mentioned above. After a follow up of 6 years the patient did not reveal any evidence of recurrence (10).

Another interesting issue touched by Sigurdson *et al.* in their paper was the potential risk of thymoma seeding that could be the negative result of a preoperative biopsy or an incomplete tumor resection in the first surgical procedure (3). This problem is sometimes raised in the literature (13-16). A lot of cases may be explained as an unfavourable progression typical for malignant neoplasms, and the iatrogenic background is questionable. However, when secondary implants were found in the needle tract after biopsy, the risk of iatrogenic tumor seeding seems to be plausible (15). Although well documented complications of surgical procedures are reported very rarely, a preoperative biopsy of mediastinal tumors is not recommended if the radiological findings indicate a resectable tumor, and suggest a thymoma. However, the probability of thymoma development in the paediatric population is very low, and usually other malignancies should be taken into consideration first thus a preoperative biopsy is justified (7).

Sigurdson *et al.* presented in a concise way a practical approach to evaluation and management of a thymoma in a young patient (3). The diagnostic and therapeutic recommendations approved for adults were analyzed and adopted for children. In light of the lack of official guidelines, established by scientific societies for paediatric oncologists, this advice given by experts of different specialities may help oncologists attending to young patients to face a very rare paediatric malignancy—a thymoma.

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