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 et 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 pediatric 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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**References**

1. Ettinger DS, Riely GJ, Akerley W, et al. Thymomas and thymic carcinomas: Clinical Practice Guidelines in
Oncology. J Natl Compr Canc Netw 2013;11:562-76.

2. Girard N, Ruffini E, Marx A, et al. Thymic epithelial tumours: ESMO Clinical Practice Guidelines for diagnosis, treatment and follow-up. Ann Oncol 2015;26 Suppl 5:v40-55.

3. Sigurdson SS, Roden AC, Marom EM, et al. Case presentation and recommendations from the April 2018 ITMIG tumor board: an international multidisciplinary team. Mediastinum 2019;3:4.

4. Stachowicz-Stencel T, Orbach D, Brecht I, et al. Thymoma and thymic carcinoma in children and adolescents: a report from the European Cooperative Study Group for Pediatric Rare Tumors (EXPeRT). Eur J Cancer 2015;51:2444-52.

5. Carretto E, Inserra A, Ferrari A, et al. Epithelial thymic tumours in paediatric age: a report from the TREP project. Orphanet J Rare Dis 2011;6:28.

6. Rod J, Orbach D, Verité C, et al. Surgical management of thymic epithelial tumors in children: lessons from the French Society of Pediatric Oncology and review of the literature. Pediatr Blood Cancer 2014;61:1910-5.

7. Gun F, Basak Erginel B, Aysegul Unuvar A, et al: Mediastinal Masses In Children: Experience With 120 Cases. Pediatr Hematol Oncol 2012;29:141-7.

8. Fonseca AL, Ozgediz DE, Christison-Lagay ER, et al. Pediatric thymomas: report of two cases and comprehensive review of the literature. Pediatr Surg Int 2014;30:275-86.

9. Saha S, Suhani S, Basak A, et al. Pediatric Thymoma with a Difference: Report of a Case and Review of Literature. J Surg Tech Case Rep 2014;6:64-6.

10. Iorio R, Evoli A, Lauriola L, et al. A B3 Type-Thymoma in a 7-Year-Old Child with Myasthenia Gravis. J Thorac Oncol 2012;7:937-8.

11. Liu T, Al-Kzayer LF, Xie X, et al. Mediastinal lesions across the age spectrum: a clinicopathological comparison between pediatric and adult patients. Oncotarget 2017;8:59845-53.

12. Travis WD, Brambilla E, Burke AP, et al. WHO classification of Tumours of the Lung, Pleura, Thymus and Heart. 4th ed. Lyon: IARC, 2015.

13. Vannucci J, Pecoriello R, Ragusa M, et al. Multiple pleuropericardial implants of thymoma after videothoracoscopic resection. Interact Cardiovasc Thorac Surg 2010;11:696-7.

14. Kattach H, Hasan S, Clelland C, et al. Seeding of stage I thymoma into the chest wall 12 years after needle biopsy. Ann Thorac Surg 2005;79:323-4.

15. Nagasaka T, Nakashima N, Nunome H. Needle tract implantation of thymoma after transthoracic needle biopsy. J Clin Pathol 1993;46:278-9.

16. Aubert A, Chaffanjon P, Brichon PY. Video-assisted extended thymectomy in patients with thymoma by lifting the sternum: is it safe? Ann Thorac Surg 2004;77:1878; author reply 1878.

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