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Malignant rhinological tumours in children

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The incidence of malignancy in children is very low, the figures for Belgium in 1980 being 12 new cases per 100,000 children per year. However, malignant tumours were, in the same statistical report, ranked as second most important cause of death in children between 1 and 15 year of age, with 11%; it was only preceded by accidents (54%).

We found it important to discuss our own results concerning malignant rhinologic tumours in children, because of the insidious start of the disease and because of the advances that are made during the last 5 to 10 years in the treatment of these malignancies.

SUBJECTS

In the period 1950-1983, 1125 children with a malignant tumour were admitted to the Children's Hospital of the State University of Ghent. Of them, 70 (6%) presented a tumour in the otorhinolaryngological area. In these figures are not included those malignancies which readily present localizations in the E.N.T.-region, such as leukemias. From these 70 children, 33 had a primary localization presenting as a neckmass, while the remaining 37 had a primary tumour in the proper otorhinolaryngological regions: ear, nose, middle ear, paranasal sinuses, nasopharynx, larynx, etc....* From the 37 children with a primary localization in the proper E.N.T.-area, 19 had a tumour in the rhinological area: 9 in the nasopharynx, 7 in the paranasal sinuses and 3 in the soft tissue of the canine fossa.

Histopathology, age and sex

Malignant non-Hodgkin lymphoma and rhabdomyosarcoma were the most common tumours. All lymphomas were classified as highly malignant and the majority were of the lymphoblastic type; we did not see well differentiated lymphomas in children. We only have one case of nasopharynx-carcinoma (NPC): it occurred in a Kaukasian boy. In other parts of the world (North-Africa, Hong Kong and

* The full history of all 37 children with a malignant otorhinolaryngological tumour is discussed in another article of the same authors (Benoit et al., 1984).

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other Asian countries) the percentage of this malignancy is much higher. From these figures we can conclude that the majority of rhinological malignancies in childhood are very aggressive tumours (Table 1). When looking at the age distri-

Table 1. Histological classification of 19 malignant rhinologic tumours in children.

	nasopharynx	paranasal sinuses	canine fossa	total
malignant lymphoma	5	3	–	8
rhabdomyosarcoma	2	1	3	6
fibrosarcoma	1	–	–	1
NPC	1	–	–	1
Ewing sarcoma	–	1	–	1
estesioneuroblastoma	–	1	–	1
osteosarcoma	–	1	–	1
	9	7	3	19

bution we see that rhabdomyosarcoma seems to occur much more frequently in younger children, while non-Hodgkin malignant lymphoma was more common in children between 6 and 12 years of age (Figure 1). Malignant rhinological tumours seem to affect more boys than girls. In our series only 3 females suffered from a rhinological malignancy (vs. 16 boys).

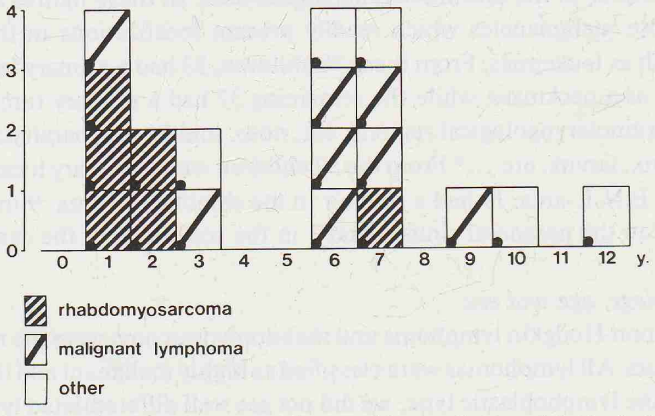

Figure 1. Age distribution in 19 children with a malignant rhinological tumour.

Early symptoms and extension

The start of rhinologic malignancies is very insidious. In most cases a painless swelling is the first or only symptom. Of course, this is only true for tumours of the soft tissue of the face and sometimes for malignancies of the paranasal sinuses;

this is not true for nasopharyngeal tumours. Nasal obstruction is one of the most common symptoms in childhood, but can also be the first symptom of a nasopharyngeal or paranasal sinus tumour; these tumours are also very often accompanied by secretory otitis media or earache, other very common symptoms in childhood. Tumours of the paranasal sinuses likely cause toothache, while ophthalmological symptoms are noted quite early in nasopharyngeal and paranasal sinuses tumours. In the latter cases, however, it is general too late to start an appropriate treatment in this stadium. Concerning the extension at diagnosis, regrettably only a minority was referred to us with a limited tumour or with only a tumour with local extension. Eight children already presented a metastasis at the time of diagnosis.

There is, however, a very close relationship between the extension at diagnosis and the survival rate. No single child with metastases survived, while of the 10 children with only local extension of the tumour, five survived, as well as the only child with a very limited tumour (Table 2).

Table 2. Extension at diagnosis and survival.

	tot. number	alive
limited tumour	1	1
local extension	10	5
metastases	8	0
	19	6*

* One of these children has a follow-up of only 30 months; the remaining 5 have a follow-up of at least 5 years and are off treatment.

Principles of treatment

It is not our intention to discuss here in detail the different ways of treatment, as they were applied in the individual children, but we will focus on some general principles. Contrary to what is seen in adulthood oncology, we always deal in pediatric oncology with very aggressive, fast growing and rapidly metastasing tumours. These tumours should consequently be considered as micro-metastasized from the beginning. Pediatric malignancies are in general very sensitive for well-defined specific antimitotic combinations, such as the L.S.A_2L_2-regimen for T-lymphoblastic lymphomas (Wallner et al., 1976), the B-cell non-Hodgkin lymphoma-regimen for B-lymphoblastic lymphomas (Hense et al., 1983; Patte et al., 1983) and the V.A.C.A.-regimen for rhabdomyosarcoma (Flamant, 1978; Tefft et al., 1977). Consequently, children presenting these malignancies are as a rule treated with chemotherapy as initial treatment. It is obvious that an exact histopathological diagnosis is very important for determining the ideal anti-tumour treatment. Histochemistry, membrane-typing and electronmicroscopy may help us in establishing the exact type of the malignancy.

Another very important factor in determining the treatment of choice is the possible occurrence of late sequelae. Extended surgery and high dose radiotherapy may, especially in the young child, cause unacceptable deformities of the face. Consequently, the whole policy in determining the treatment of choice is, without any doubt, the result of a teamwork involving different specialists.

Survival

The prognosis in children with malignant lymphoma and rhabdomyosarcoma has improved markedly in recent years. Figure 2 gives us the survival rate of all children that were seen in our Children's Hospital and who presented these kinds of tumours, and not only with an otorhinolaryngological localization.

The survival rate in children with rhabdomyosarcoma during the period 1967–1969 was very poor: only 15% survival after 7 years, while the results of the most recent period 1977–1982 show a survival rate of 66% after 80 months.

Similar results are noted with regard to the survival rate in children with malignant non-Hodgkin lymphoma. Before 1974 the survival rate was 25%, 40 months after diagnosis, while it is now 60% after 5 years, and 55% after 8 years.

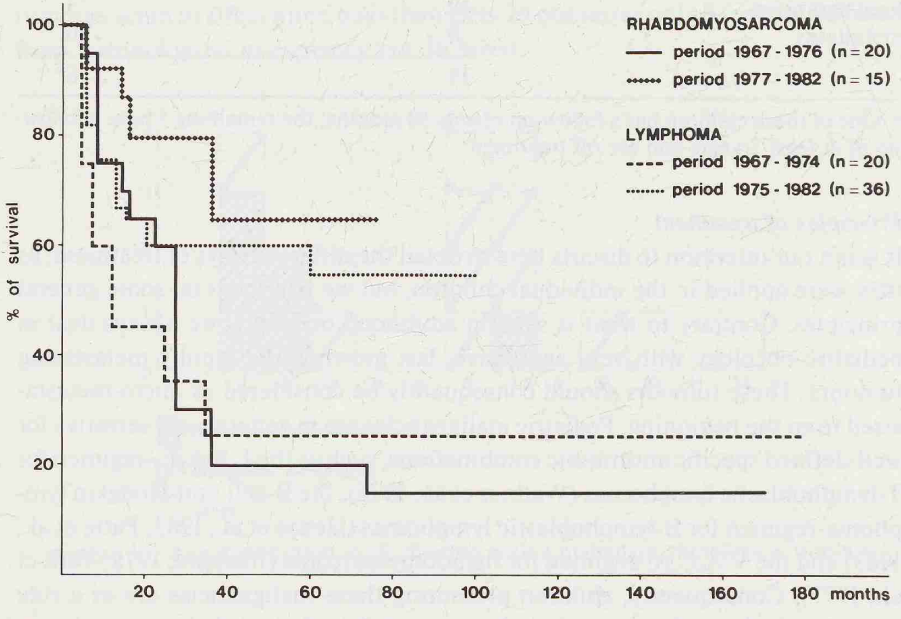

Figure 2. Survival rate in children with non-Hodgkin malignant lymphoma and rhabdomyosarcoma treated in the Children's Hospital of the State University of Ghent, according to the different time periods studied.
All localizations of these tumours are considered, and not only the otorhinolaryngological ones. (with permission of Acta oto-rhino-laryng. Belg.).

CONCLUSION

In conclusion, we can state that the early symptomatology of a malignant tumour in the rhinological area is very often insidious. Very often only a painless swelling is noted as the first and only symptom. In these cases the tumour may go unnoticed for several months. Consequently, biopsy is recommended in all cases of painless (and also painful) swelling, where an infection of benign pathology is not obvious. Children presenting secretory otitis media or nasal obstruction should always be examined very carefully and children with any neurological symptom should be examined properly to exclude a malignancy. The results of the treatment have been improved considerably in recent years thanks to the new chemotherapy regimens, so that extensive surgery is not recommended in these children and that radiotherapy may also be avoided.

REFERENCES

1. Benoit Y, Delbeke MJ, Cauwenberge P van, Kluyskens P. Malignant otolaryngological tumours in children. Acta Oto-rhino-lar Belg 1984; 38:288.
2. Flamant F. Rhabdomyosarcoma Group SIOP therapeutic trial. SIOP X Annual Conference, Abstracts, 1978; 15
3. Hense G, Langermann HJ, Muller-Weirich S, Riehm H. Different approaches to the treatment of childhood B-cell lymphomas. SIOP XV Annual Conference, Abstracts, 1983; 154.
4. Patte C, Philips T, Bernard A et al. Updated results of the French protocol for B-cell non-Hodgkin lymphoma on ninety patients. SIOP XV Annual Conference, Abstracts, 1983; 111.
5. Tefft M, Fernandez CH, Moon TE. Rhabdomyosarcoma response with chemotherapy prior to radiation in patients with gross residual disease. Cancer 1977; 39:665.
6. Wallner M, Biercheral JM, Lieberman PH et al. Non-Hodgkin lymphoma in children: a complete study of two modalities of therapy. Cancer 1976; 37:123.

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