



Original article

Epidemiological Consideration About Pediatric Sarcomas in Albania

Donjeta Bali (Alia)¹, Fabian Cenko², Anila Godo³, Eleni Nastas⁴, Mirela Xhafa⁵, Numila Maliqari⁶, Lila Shundi⁷

¹Service of Oncohematology, Pediatric Department, University Hospital center "Mother Theresa"-Tirana Albania

² Catholic Medical University "Our Lady of Good Council"-Tirana, Albania

^{3,4&5} Service of Oncohematology, Pediatric Department, University Hospital center "Mother Theresa"-Tirana Albania

⁶ General Pediatric Service, University Hospital center "Mother Theresa"-Tirana Albania

⁷ Public Health Institute

ABSTRACT

Background: Sarcomas are frequent in the pediatric age, occupying the fourth place (by about 10%). Objective of the present study is to present some epidemiological data of Pediatric Sarcoma (PS) in Albania, from 1997 to 2010. **Method:** Among 93 children of 0 to 14 years old with PS, we studied the specific weight of the PS in the general pediatric morbidity, vs. other pediatric cancers; annual incidence in the pediatric population, the incidence during the performance period under study; geographical distribution according to three areas of Albania, in 12 regions and 36 districts. **Results:** Sarcomas in our country constitute about 8.3% of all pediatric cancers (CI from 1.5% to 15.1%). The annual incidence in the general population is 0.19, the cumulative incidence for the period 14 years in the pediatric population is 9.1/105 children born alive, with an average annual incidence of 0.65/105 children born alive. However, within the period of study 04-10 incidence has increased almost by 50% compared with the first 7-year (0.96 vs. 0.43). High incidence was found in northern Albania, in the areas of Dibra, Kukes, Durres etc. **Conclusions:** High numbers of PS, high mortality, is forcing attention from health institutions. Dynamics study of incidence for the whole period draws attention to a growing incidence of lightly progressive; a higher incidence for years 04-10, there may be incentives in search of various, favorable, explosive, environmental or other factors that may have influence on this phenomenon.

KEYWORDS: Sarcomas, children, epidemiological data.

INTRODUCTION

Currently, neoplasm of pediatric age in Albania, according to Institute of Statistics (INSTAT) and Ministry of Health (MSH), are in 5th place overall morbidity pediatric (with about 10% of cases) and they ranked after pulmonary and neonate pathologies, as congenital anomalies. One of the most frequent cancers in children is the Sarcoma, a malignant tumor with origin from mesenchyme cellules. The aim of our study was to make a presentation of some demographic and epidemiological data of Pediatric Sarcomas (PS) in Albania, for a period of study, about 14 years, from 1997 to 2010. While the specific objectives of this study are the following: 1) to document the results about the dissemination of sarcomas in pediatric age group in

Albania; 2) to assess national incidence rate of sarcomas and compare it with international references; and 3) to identify risk factors for sarcomas in relation to epidemiological, demographic and/or geographical variables.

MATERIALS AND METHODS

This study is prospective (for the period following January 1st, 2007) and retrospective (for the period from January 1997 to December 2006). Descriptive and exploratory-through the summary and detailed description of all the cases presented in the service of Onco-Hematology, according to several variables (demographic, geographic related to place of residence, etc.) Our goal is to know the

common patterns of expression of these pathologies in the pediatric age. Also, in this way we evaluate the level of expression of sarcoma in the pediatric age group and in the total morbidity in this group age. Analytic-hereby express statistical estimation study of some variables of the disease in the context of their contribution to the emergence of the disease and its prognosis. In this way we tried to present the highest level of observer epidemiological study.

The study consists of 93 cases of pediatric age (0 to 14 years) with different types of sarcoma, diagnosed, treated and/or followed by: Pediatric Oncohematology Service, Adult Oncology Service and Surgery and Radiotherapy of

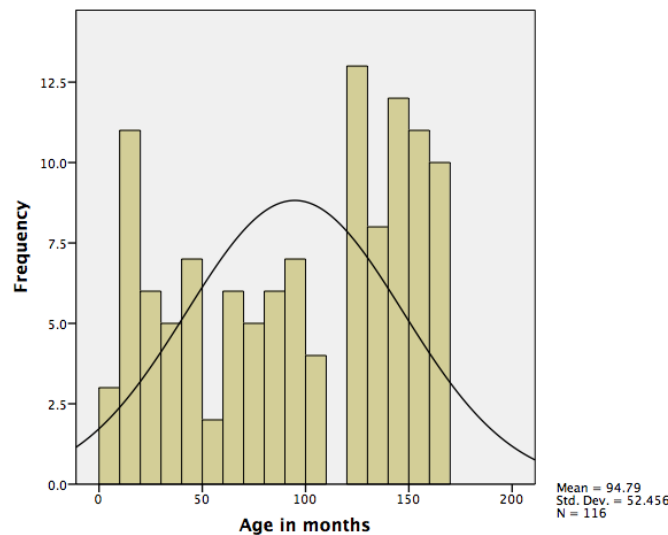
Oncology; all located in UHC "Mother Theresa" in Tirana, Albania.

Some cases (22 cases) were followed jointly with Pediatric Oncology Services in other countries such as: Italy (11 cases), Turkey (6 cases), Austria (2 cases), and Greece, Israel and Germany (1 case for each country). The table 1 describes the sample according to the variables of central tendency and distribution (expressed in months). The following chart shows the graphical distribution of our pediatric sample under study, which seems to be slightly left skewed according to Gaussian distribution (Graph1).

Table 1: The variables of central tendencies and distribution (expressed in months)

No Cases	Mean	Median	Mode	Standard Deviation	Value interval	
					Minimal value	Maximal value
93	99	108	144	52.6	1	168

Graph 1: The age-based distribution of the sample compared with normal distribution (Gaussian)



The criteria to include in the study were the age at the time of diagnosis: 0-14 years old, the histopathology confirmation of sarcoma and residents in Albania. In the early years of the study, when the Pediatric Oncology Service was transferred to the Pediatric Department, some cases continued to be followed and treated in the Department of Adult Oncology (7 cases).

We have full rights to say that our data are given as the national character because the services of Pediatric Onco-Hematology, of Adults, of Surgery and Radiotherapy, are unique in the tertiary system of organization of health care, so every child with pediatric sarcoma must be followed by those services. The study parameters analyzed were: the specific weight of pediatric sarcoma in our country in general pediatric morbidity, in neoplasms of the pediatric age.

Average incidence annual /10⁵ of: the population as a whole, the pediatric population in particular, for sarcoma and its 4 main types.

The performance of this incidence: for the whole period in our research. (1997-2010), compared between the 2 periods

of 7 years (1997 to 2003 and 2004 to 2010) and the entire time period.

Geographical distribution: We analyzed the distribution of sarcoma cases by 12 regions of the country (Regions), 36 districts, in the northern, central and south of the country.

Regional geographical distribution was practiced based on the possibility of the type or proximity within these areas to different environmental factors of socio-economic level, living habits, the way of nutrition, etc. which may play a role in the etiology of sarcoma. The aim was to investigate the possibility of the influence of these factors on the incidence, clinical forms, the gravity of the disease. Thus, removing the influence of the factors mentioned above in our data, we minimize the maximum relativity results and increase their accuracy. For all the above parameters were use demographic data from Statistical Department (INSTAT) 2003, including the Statistical Department in the Ministry of Health, for the years of study on the structure of pediatric morbidity.

The original row data initially were organized in excel database and in a later phase was imported to SPSS program

in order to carry out the statistical analyses ("The Statistical Package for the Social Sciences", version 13.0 - SPSS, Chicago, Illinois, USA). Correlation between continuous variables were tested through the student test, while for categorical variables it was applied the chi square test (χ^2). The threshold value for determining the statistical tests of significances was fixed at p value ≤ 0.05 , where $p \leq 0.05$ was considered statistically valid correlation and for values of $p > 0.05$ correlation is not statistically valid.

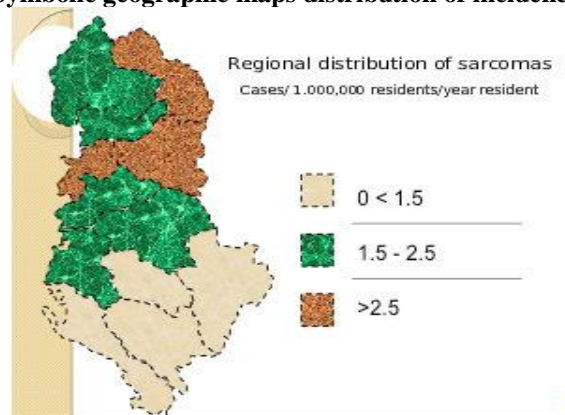
RESULTS

The table 2 shows the number of new cases, diagnosed with sarcoma, for each year of the study period (1997-2010), and the incidence rate for the live birth cohort and annual incidence for pediatric age-group (0 – 14 years). In order to understand better the dynamics of the average annual incidence of the pediatric sarcoma in our country for our period of study we have presented this graph (graph 2).

Table 2: The distribution of cases and annually incidence for each year during 1997-2010

Year	No. of births	No.of new cases	Incidence (for 10^5 baby birth live)	Annual Incidence (per 10^5 children0 – 14 years)
1997	99,584	4	4.0	0.4
1998	97,003	9	9.3	0.9
1999	93,469	11	11.8	1.1
2000	82,653	1	1.2	0.1
2001	87,558	2	2.3	0.2
2002	73,415	4	5.4	0.4
2003	75,830	6	7.9	0.6
2004	69,394	8	11.5	0.8
2005	63,894	5	7.8	0.5
2006	55,211	10	18.1	1.0
2007	53,491	9	16.8	0.9
2008	58,472	8	13.7	0.8
2009	58,472	9	15.4	0.9
2010	58,472	7	12.0	0.7
1997 - 2010	1,026,919	93	9.1	0.65

Graph2 : Symbolic geographic maps distribution of incidence by region



The occurrence of new cases of sarcomas per year is presented together (In the table 3) with its 4 constituent types (OS, ES, RMS dhe NRSTS). RMS and ES show a increasing trend in the second half of the period under study

(2003 – 2010), while the incidence of new cases with OS and NRSTS doesn't change. The average annual incidence of soft tissue sarcoma (STSs) in our study is 3.9/ 1,000,000 children or 1.2/1,000,000 inhabitants. The average annual

incidence resulted RMS 2.6 /million children, while annual incidence NRSTS is 1.3/million children. For bone sarcoma (BS) resulted incidence 2.6/million children. Our annual incidence for OS resulted in 1.2/ million children, while for

ES is 1.4/million. Our national data doesn't comply with international bibliography on this argument since for the last 30 years the trend of ES stays stable.

Table 3: Yearly based distribution of new cases of sarcoma, and its four subtypes, according to the study period

Year of diagnose	Different subtypes of sarcoma				Total sarcoma
	Osteosarcoma	Ewing	RMS	NRSTS	
1997	2	0	1	1	4
1998	0	1	7	1	9
1999	1	1	4	5	11
2000	0	0	1	0	1
2001	0	0	2	0	2
2002	0	1	0	3	4
2003	1	0	4	1	6
2004	3	4	0	1	8
2005	0	1	3	1	5
2006	3	4	2	1	10
2007	2	2	5	0	9
2008	2	2	3	1	8
2009	3	2	3	1	9
2010	0	2	3	2	7
Total no	17	20	38	18	93

There are few cases of sarcoma diagnosed each year in Albania which would make not reliable the yearly-based analyses. Therefore the study period was divided into two equal and consecutive time intervals (1997 - 2003 and 2004 - 2010). According to this division, the Table 4, shows the cumulative number of cases and risk of developing sarcomas for along each time interval. So, the risk to develop a sarcoma for a child aged 0 – 14 years old during

the second period of the study period (2004 - 2010), is doubled compared with the first one (RR. 1.5 vs. 0.7). Incidence for the period 14 years is calculated on the basis of the pediatric population (0-14 years) of 1,026,919 inhabitants; while for the periods 1997 to 2003 and from 2004 to 2010 the estimated pediatric populations are 1,219,024 and 834.812 (estimates were made according to INSTATs for these periods).

Table 4: Comparative incidence of sarcomas amongst two 7-years interval, composing the study period

Periods	New cases	Incid. of period /10 ⁵ children (0-14 years)	Annual Incid. /10 ⁵ children (0-14)	RR
7 years ('97-'03)	37	3.04	0.43	0.7
7 years ('04-'10)	56	6.71	0.96	1.5
14 years ('97-'10)	93	9.06	0.65	1

The table 5, shows the cumulative distribution of cases, diagnosed during the study period, for each district, which was expressed as incidence rate as well (to be noted that in this table the denominator expresses each district total population and not just pediatric-age population).

The Table 6 shows the regional based distribution of number of cases and annual incidence relative to 14 years study interval. The data obtained from Table 5, help identify regions with higher incidence rate, as compared with the national average annual incidence. According to these data the regions with the highest incidence rate are Durres

Kukes, Dibra, Elbasan and Lezha. In the graph 2 are presented regional-based average incidence rates, which are clustered in three groups in order to evidence the geographical areas with low (beige), medium (green) and high risks (brown) for pediatric cases of sarcoma according to national incidence value of 2.5 cases / 1,000,000 total population. Table 4, expresses the dynamics of the number of cases (which represent a good indicator of incidence) within the 14 years study period divided into two 7-years intervals. In most of the regions it is noticed an increasing trend relative to the second half interval.

Table 5 : Distribution of new cases and annual average incidence by districts

Districts	Population	Cases/14 year	Inc./10 ⁵ abitants/year
Berat	148050	4	0.19
Bulqize	47842	0	0
Delvine	32561	0	0
Devoll	39562	0	0
Diber	103614	6	0.41
Durres	172013	11	0.46
Elbasan	222972	6	0.19
Fier	212318	9	0.30
Gramsh	47245	2	0.30
Gjirokaster	66394	1	0.11
Has	23789	0	0
Kavaje	95427	0	0
Kolonje	28356	0	0
Korce	194258	7	0.26
Kruje	64211	1	0.11
Kucove	41918	0	0
Kukes	88321	5	0.40
Lac	55414	0	0
Lezhe	67192	3	0.32
Librazhd	79750	4	0.36
Lushnje	140287	1	0.05
M.Madhe	49410	0	0
Mallakast.	45568	0	0
Mat	82608	3	0.26
Mirdite	48869	3	0.44
Peqin	34293	0	0
Permet	41741	0	0
Pogradec	80211	0	0
Puka	52922	1	0.13
Saranda	59169	0	0
Skrapar	45584	2	0.31
Shkodra	206717	6	0.21
Tepelena	51907	0	0
Tirana	414367	16	0.28
Tropoja	49054	1	0.15
Vlora	189151	1	0.04

Total	3.465.384	93	0.19
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Table 6: Distribution of cases and annual average incidence by regions for the period

No.	Regions	Population	Cases /14 year	INC./10 ⁵ abitants /year
1.	BERAT	278,317	6	0.15
2.	DIBËR	234,064	9	0.27
3.	DURRËS	236,224	12	0.36
4.	ELBASAN	349,967	12	0.25
5.	FIER	398,173	10	0.18
6.	GJIROKASTËR	160,042	1	0.04
7.	KORÇË	342,387	7	0.15
8.	KUKËS	161,168	6	0.27
9.	LEZHË	171,475	6	0.25
10.	SHKODËR	309,049	7	0.16
11.	TIRANË	544,087	16	0.21
12.	VLORË	280,881	1	0.03
	TOTALI	3,465,834	93	0.19

DISCUSSION

The incidence's performance during the period of all 14 years, shows that in its second half there is a clear upward trend which can be explained as objective reasons (real growth in the number of new cases) or reducing the rate of population growth, as well as those subjective (improvement of diagnostic capacity at the National Pediatric Oncohematology Service).

In particular for STSs, at international level, a progressive increase significantly ($p < 0.05$) by about 1% per annum of the cases referred to in the literature [3].

Regarding incidences for each group or separate species we found too, that the average annual incidence of Soft Tissue Sarcoma (STSs) is 3.9/1,000,000 children or 1.2/1,000,000 inhabitants. Different authors quote figures that vary from 3/million in Asia, up to 12 cases / million in North America[4,11], others give an international incidence of 11/million [5,12]; while for RMS annual incidence with average resulted in 2.6 million children, the same authors refer to an incidence that varies from 4 to 6/million children [1,5]; for NRSTS 1, 3/million children, while the world's refers about 5/million [1]

For bone sarcoma our incidence resulted in 2.6 / million children, while in contemporary literature get referred from 2-8 / million children aged 0-14 years [6] and for U.S. refers 7/million [2] for OS incidence our resulting annual 1.2 / million children, while for ES is 1.4 / million children. Literature data on the annual incidence of these two entities

refers to respectively 3.7 and 2.1 / million / children less than 15 years [2, 9]. As seen from our data, ES find with frequency OS, but always within the figures referred to the global data [10, 7, 8].

The study of geographical distribution, arise a high incidence in some regions of the country, such as a Durres, Diber, Kukes, Elbasan and Lezha.

Despite the growing data of the sarcoma's incidence in our country in the last 7-years-old, their remain within the limits of those referred to in contemporary world literature for sarcoma in general as well as two large groups or specific types of their constituent [8].

CONCLUSION

The sarcomas, ranked the fourth place of the list, between the most frequent neoplastic pathologies of children, in our study. The results give a national annual average incidence around 2/million inhabitants of the population, the annual average incidence of sarcoma is 6.5/million inhabitants' paediatric population. Incidence of our period of study is: 91/million paediatric population. Dynamic study in both 7-years periods provides interesting data, with statistically significant changes, a statistically higher incidence for the year 2004-2010, that can be catalytic in search of Various ,favorable, explosive, environmental or other factors that may have an influence on this phenomenon. In the northeastern areas, the incidence is higher.

We have no comments, but we want to draw attention to the digits, suggesting multilevel studies in search of influencing factors or predisposing situations. There are digits that taking into account the still high mortality of this pathology in children oblige attention from institutions of diagnostic-curative health services at all levels, in order to complete the field of politics of public health. We must design as soon as possible a national registry for paediatric neoplasms in general and in particular for the sarcomas.

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*Corresponding author: Donjeta Alia(Bali)
E-Mail: donjetabali@yahoo.com