Original Article

Mediastinal and Chest Wall Masses in Children: A Single Institution Study

Mitra Mehrazma[1], Soraya Salehi[2], Shokrollah Yousefi[1], Selaheddin Delshad[3], Ahmad Jalilvand[1], Alireza Hasanpour[1]

1. Department of Pathology, School of Medicine, Iran University of Medical Sciences, Tehran, Iran.
2. Department of Pediatrics, School of Medicine, Iran University of Medical Sciences, Tehran, Iran.
3. Department of Surgery, School of Medicine, Iran University of Medical Sciences, Tehran, Iran.

ABSTRACT

Background and Objective: **Mediastinal masses in children are a heterogenous group of asymptomatic potentially life-threatening congenital, infectious, or neoplastic lesions that present complex diagnostic and therapeutic dilemmas.**

Materials and Methods: **The clinical and laboratory features of 65 patients who had open biopsies in Ali-Asghar hospital over 11 years were reviewed.**

Results: **Seventy-two percent of cases were male and 28% were female. The age varied between 2.5 months and 19 years. Eighty-four percent of lesions were malignant and 16% were benign. Neurogenic tumors were the most common (36.9%). In order of frequency the following lesions were the most common neuroblastoma (mostly in posterior mediastinum), Hodgkin's and non-Hodgkin's lymphoma (mostly in middle mediastinum), and malignant small round cell tumor (mostly in chest wall). Most patients were presented by fever (21.5%), dyspnea (20%) and cough (12%), especially in Hodgkin's and non-Hodgkin's lymphoma. Lymphadenopathy (29.2%) and hepatosplenomegaly (13.8%) were the most frequent clinical signs. Laboratory examination revealed anemia (29%), leukocytosis (35%), and high sedimentation rate (21.5%).**

Conclusion: **The clinical presentation and laboratory findings of mediastinal and chest wall masses are often non-specific and are variable, but according to their clinical data (i.e. age) and location, only few important diagnoses should be considered.**

Key words: **Mediastinal, Chest wall, Mass, Children**

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Address communications to: Dr. Mitra Mehrazma ,Department of Pathology, Ali-Asghar Hospital,Dastjerdi St., Tehran- IRAN
E-mail: mitmehr@yahoo.com

Introduction

Chest wall masses at childhood have often a mediastinal origin and comprised of a wide range of congenital cysts and dense malignant and cystic tumors (1). Urgent cases with obstructive respiratory symptoms are a frequent manifestation in these lesions due to their special site of involvement and narrow airways in children. The incidence rate of malignant mediastinal tumors in children is 40-72% and due to its risk and high arte of respiratory failure, its fast diagnosis is very essential (2).

Mediastinal masses with regard to their origin are categorized into anterior, middle, and posterior types. The common forms of anterior type include lymphomas, thymus masses, teratoma, angioma, lipoma, and thyroid masses. Middle mediastinal type comprised lymphoma, metastatic lesions, infectious diseases with involvement of lymph nodes, pericardiac cysts, bronchogenic cysts, esophageal lesions, and hernias. The most prevalent form of posterior masses is neurogenic tumors. In addition, posterior masses include 20% of primary mediastinal masses and cysts. Other kinds of posterior mediastinal masses consist of enterogenic cysts, thoracic hernias, and malignant tumors such as Ewing's sarcoma, lymphoma, and rhabdomyosarcoma, and leukemic masses. Dyspnea and a history of breathing difficulties may indicate a lesion which imparts pressure on airways. In this respect, weight loss, nocturnal perspiration, and general weakness and malaise should also be considered. Exact physical examination may show a pressure on superior vena cava which expresses itself as facial redness and vasodilatation. Sometimes, masses above clavicle and neck may show themselves as firmness and swelling (3-4).

Chest radiography, CT-scanning with venous contrast, and MRI are helpful in diagnosis and localization of tumor. Final diagnosis in all cases is only possible through biopsy and histological evaluation (5-6). The most common diagnosis for mediastinal masses using histological methods is tumors which contain round small cells and include Ewing's sarcoma, primary neuroendocrine tumor (PNET), rhabdosarcoma, lymphoma, and leukemic masses. Immunohistochemical and electron microscopic techniques are required for their final identification (7-9).

Therefore, this study was conducted to perform a 10 years study on mediastinal masses, their location, symptoms, clinical and paraclinical findings, and their prognosis in Ali-Asghar children hospital as a reference center for tumors of children in Iran. The results of this study can be useful in differential diagnosis of such tumors for physicians and pathologists and this can greatly lead to prompt and early treatment of the condition

Materials and Methods

The descriptive retrospective strategy of this study was performed on the past 10-year referrals of Ali-Asghar children hospital with a diagnosis of mediastinal and chest wall mass who underwent surgical operation and/or after operation again referred to the hospital for further treatments and their biopsies were evaluated as obtained from recording office of pathology section and their samples were re-evaluated separately by two pathologists as necessary.

In addition, their clinical, radiological, paraclinical, and follow-up records were collected and analyzed. For statistical analysis, SPSS software was used and a p value less than 0.05 considered significant.

Results

In this study, a total of 65 cases (72% and 28% of them were boys and girls respectively) were studied. Their age range was from 2.5 months to 19 years. The most common year for the disease incidence was year 2 and the mean age was 7 years. In addition, 84% and 16% of the tumors were malignant and benign in nature respectively. Meanwhile, incidence rate for posterior, middle, anterior mediastinal, and chest wall masses was 34.1, 29.3, 22, and 12% respectively (Table 1).

Table 1. Incidence rate of thoracic masses in referrals of Ali-Asghar children hospital

Chest wall	Posterior mediastinum	Anterior mediastinum	Middle mediastinum and chest wall
Round small cell tumor 3	Neuroblastoma 6	Malignant lymphoma 3	Hodgkin's lymphoma 1+5
Malignant lymphoma 1	Other neural tumors 3	Hodgkin's lymphoma 2	Malignant lymphoma 1+1
	Primary neuroendocrine tumors 3	Cyst 2	Neuroblastoma 2
	Malignant tumor of peripheral nerve sheath 1	Round small cell tumor 1	Round small cell tumor 2
			Reactive masses 1

All cases of neuroblastoma and the majority of Hodgkin's cases had an age below 10 years and above 8 years (with an exception of two cases) respectively. In addition, cases of malignant lymphoma had an even-distributed age. For other mediastinal masses, no significant correlation was found out with regard to age. Table 2 shows mediastinal masses with regard to percentage, number, and site of involvement.

Table 2. Causes of mediastinal masses with regard to prevalence and the most common site of involvement in cases of Ali-Asghar children hospital

Diagnosis for mediastinal masses	Number	Percentage	The most common site of involvement
Neuroblastoma	14	21.5	Posterior mediastinum
Hodgkin's lymphoma	13	20	Middle mediastinum
Non-Hodgkin's lymphoma	11	16.9%	Anterior mediastinum
Round small cell tumor	6	9.2%	Chest wall
Neural tumors (except for neuroblastoma)	5	7.7	Posterior mediastinum (only)
Primary neuroendocrine tumor	3	4.6%	Posterior mediastinum (only)
Cyst	2	3.1%	Anterior mediastinum (only)
Malignant tumor of peripheral nerve sheath	2	3.1%	Posterior mediastinum (only)
Reactive masses	1	1.5%	Middle mediastinum (only)
No diagnosis	4	6.2%	Middle mediastinum (only)
Other diagnoses	4	6.2%	Middle mediastinum (only)
Total	65	100%	

Furthermore, non-Hodgkin's lymphomas were mostly lymphoblastic, Burkitt's, B-cell and giant cell, and anaplastic large cell in nature in order of frequency. Meanwhile, Hodgkin's lymphoma cases were mainly nodular sclerosing and then a mixed cellularity in nature. The most common symptoms in patients were general malaise (24.6%), fever (21.5%), dyspnea (20%), and coughing (12%). In this respect, Hodgkin's and non-Hodgkin's lymphomas were the most common causes of these symptoms. The most common site of these symptoms was masses within middle mediastinum. The patients had these symptoms from several days to a maximum of 36 months (with an average of 6 months). Meanwhile, most of patients referred for diagnosis and treatment within one month.

The most common symptoms in examined patients were lymphadenopathy (29.2%), splenomegaly (13.8%), and hepatomegaly (13.8%) and Hodgkin's and non-Hodgkin's lymphoma and small round-cell tumor were the most prevalent conditions with the presence of all of these symptoms. Laboratory analysis showed that 29% of patients had anemia, 35% had leukocytosis, and

12.3% had lymphocytosis. In addition, platelets were normal in 30.8% of cases and thrombocytosis was observed in 6.2% of cases. Increased level of ESR was observed in 12.3% of cases. Meanwhile, anemia, leukocytosis, thrombocytosis, and increased ESR were more accompanied with Hodgkin's lymphoma than other conditions.

Discussion

The results of this study showed that the incidence of malignant tumors is nearly 5 times that of benign ones and with regard to existing data, neuroblastoma, Hodgkin's and non-Hodgkin's lymphoma are the most prevalent mediastinal tumors. In this regard, neuroblastoma and lymphomas are more common at lower and higher age respectively. In addition, the most common site of involvement was posterior mediastinum. The results of this study was nearly similar to a study by Grossfeld et al on 196 children as referrals of surgery section of Indiana school of medicine in that the rate of malignant tumors is more than threefold as compared to benign and similar tumors (1). Meanwhile, the results of our study were similar to a study by Campbell and Simpson on 121 referrals of Melbourne children hospital (10). In addition, Massie et al (Australia) showed that malignant mediastinal tumors in children are more common than benign ones and the most common form is neurogenic tumors (11). In another study by Sairanen et al, it was found out that the rate of incidence of benign tumors is slightly greater than malignant ones (12). The main reason for this difference with regard to our results can be attributed to those patients as referrals of chemotherapy.

The location of mediastinal masses and their prevalence with regard to anterior, middle, and posterior portions is consistent with anatomical, histological, and embryological characteristics of these regions. As mentioned before, mediastinal space has mesothelial surfaces and contains various vital organs including heart, large vessels, phrenic and vagus nerves, trachea, bronchi, esophagus, lymph nodes, and thymus. Although some authors divide mediastinum into 4 sections, but others prefer simpler Felson classification system on the basis of radiological hallmarks. In the latter case, mediastinum is divided into three sections, i.e. anterior, middle, and posterior. This classification has a good relationship with the pathogenic processes of some disorders.

In general, masses of anterior mediastinum include thymus tumor, germinal cell neoplasms, lymph node proliferation (Hodgkin's and non-Hodgkin's lymphoma, reactive lymphadenopathy, and metastases), and ectopic lesions of thyroid and parathyroid. Lesions of lymph nodes, soft tissue tumors, foregut cysts, and aneurisms are located within middle mediastinum. The common tumors of posterior mediastinum include neural malignancies, soft tissue tumors, and aneurisms (13).

The initial symptoms as a reason for refer is impart of pressure on airways, upper thorax, and large veins, general weakness, and fatigue, especially at the first few months. King et al indicated that these symptoms are more pronounced in children below 2 years (14). Massie showed that these symptoms are helpful for localization of the tumor (11). In our study, most of the patients referred to physician within one month that can explain the shortness of symptoms.

The important factors for differential diagnosis of mediastinal tumors are age, gender, symptoms, and history. There are also little clinical symptoms for localization of the tumor. If tumor reaches a significant size, due to its pressure, such symptoms like chest pain, dyspnea, dysphagia, coughing, heart failure, and impact of pressure on superior vena cava may appear. Except for the latter two symptoms, the other ones do not help in correct differential diagnosis of malignancy. Therefore, surgical operation and pathological examination are required for definite diagnosis (15). Meanwhile, all of the paraclinical findings are non-specific and can not help in diagnosis. It is noteworthy that in case of a larger population and long-term follow-up, it is possible to find a relationship between some factors like ESR and the presence of some disorders.

Conclusion

Small round cell tumors were categorized into a unified group in this retrospective study and further immunohistochemical studies are required to determine their type.

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