Pediatr Radiol (1986) 16: 240-244

Pediatric Radiology

Primary leiomyosarcoma of the lung in a girl

G. Beluffi[1,2], P. Bertolotti[1], A. Mietta[1], G. Manara[3] and M. Luisetti[3]

[1]Department of Radiodiagnosis, [2]Section of Pediatric Radiodiagnosis, IRCCS Policlinico "S. Matteo" and [3]Institute of Phthisiology and Pulmonary Diseases, University of Pavia, Pavia, Italy

Abstract. Leiomyosarcoma of the lung is a type of tumour rarely found in childhood. It arises from smooth muscle either of bronchial or arterial walls, has a variable pattern of local growth, blood-borne metastatic spread with lymph nodes sparing and a clinical course characterized by fever, cough and worsening dyspnea. We report a case which occurred in a 14-year-old girl, reviewing all the other cases found in the literature.

Primary leiomyosarcoma of the lung is a malignant tumour occurring rarely both in children [2-5, 6, 8, 9, 11] and adults [1, 2, 7, 10].

The tumour arises from smooth muscle either of the bronchial or arterial walls and is characterised by a variable pattern of local growth and blood-borne metastatic spread, while lymphatics and lymph nodes are usually spared [2, 7, 10].

Since to our knowledge only nine cases of this tumour have been reported in the literature [2, 4-6, 8, 9, 11], we present this case in a young girl studied in our hospital.

Case history

This girl was 14 years old when she first came to our observation. Her illness had begun 9 months previously with fever, cough and chest pain: a chest film performed at that time showed perihilar consolidation on the right with a pleural effusion. The child was tuberculin negative and over the next months was treated with various antibiotics as well as antimycotics with a poor response. Increase in the pleural effusion and repeated episodes of hemoptysis led to hospitalization in another institution where no diagnosis was made.

At the time of admission to our hospital, chest films showed retraction of the right hemithorax and elevation of the right diaphragm with a large, ill-defined, triangular opacity on the right (Fig. 1 a).

Tomography was performed, revealing more clearly the contours: the mass was triangular in shape with a retracted appearance and localized to the right middle lobe with a small pleural effusion persisting on the right (Fig. 1 b).

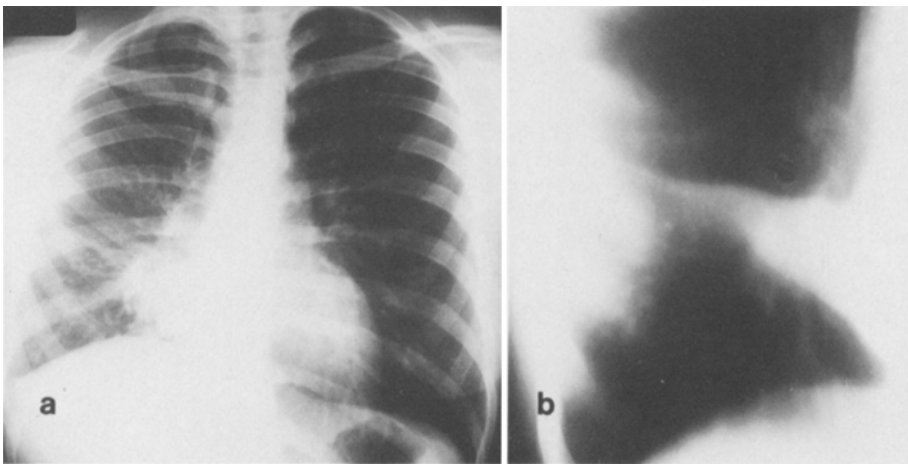

Fig. 1. **a** Retraction of the right hemithorax; elevation of the right diaphragm; ill-defined right opacity; small right pleural effusion. **b** (tomography) triangular retracted shape of the opacity localised to the right middle lobe

Table 1. Previously reported cases of lung leiomyosarcoma in children

Authors	Sex	Age	Symptomatology	X-ray findings	Localization	Bronchoscopy	Bronchography	Metastases	Treatment			Results
									Surgery	Radiotherapy	Chemotherapy	
Holinger et al. (1950)	F	5	Increasing respiratory stridor since 1½ yrs of age	Mass in the tracheal lumen	Anterior wall of the tradea (2nd to 5th ring)	–	–	None	Resection	–	–	Alive 4½ yrs after resection
	F	6	Pneumonias + increasing respiratory difficulty since 5 yrs of age; weight loss	Obstructive emphysema due to right main bronchus partial obstruction	Right main-stem bronchus	Thacheal dilatation. Friable mass extending from the carina into both bronchi	Stenosis right main bronchus	None	Unresectable	1250 R	–	Died after a course of roentgenray therapy to the tumour region for untreatable anemia
Killingworth et al. (1953)	F	7	"Smothering spells" weight loss; pain left chest	Atelectasis of the left lung	Left main-stem bronchus	Mass in the left main bronchus (non specific granulomatous inflammation)	Inadequate to draw diagnosis.	Extension of the tumour into the atrium via pulmonary vein. No extension into the hilum. No metastases in the mediastinal nodes.	Left pneumonectomy	–	–	Well and alive 2 yrs after operation
Watson and Anlyan (1954)	M	4	Cough; increasing dyspnea; hemoptysis	Tumour in the right main bronchus. Marked emphysema of the left lung	Lateral wall of the right main-stem bronchus	Tumour in the right main bronchus	–	None at necropsy	Thoracotomy: attempted removal	–	–	Died during operation for cardiac arrest
Merritt and Parker (1957)	F	8	Cough; retrosternal pains; weight loss; vomit; dyspnea and wheezing	Tumour left upper lobe, posteriorly to the mediastinum	Left main bronchus (at the level of the upper lobe orifice)	Narrowing of the main bronchus. Biopsy unsuccessful	–	None	Lobectomy	–	–	Alive 5½ yrs after operation
Lawson and Goldstein (1971)	M	6	Sudden onset of cough and fever following tonsillectomy	Atelectasis of the left lung	Just distal the carina into upper and lower left second and third order bronchi	Obstructing tissue mass in the left main-stem bronchus	–	None	Left pneumonectomy	–	–	Asymptomatic

Table 1 (continued)

Guccion and Rosen (1972)	F	11	?	Mass right upper lobe	Right upper lobe	?	?	None	Resection	?	?	Alive 31 yrs after operation
	M	new-born	Respiratory distress	Mass left upper lobe	Left upper lobe	?	?	None	–	–	–	Died shortly after birth
Ownby et al. (1976)	M	1.2	Fever; decreased appetite; noisy, labored respiration	Large mass in the right upper lobe	Posterior segment of the right upper lobe	–	–	Upper part of the right lung (4 mths after resection of the tumour). Right posterior cerebral fossa mass (8 mths after tumour resection)	Right upper lobe resection	3050 + 1010 R (to recurrence in the upper part of the right hemithorax) 2900 R (to the right posterior fossa recurrenoe)	Vincristine sulfate + cyclophosphamide	Died 15 mths after first operation (death 1 mth after posterior fossa surgery due to acute hydrocephalus. Residual tumour found in the brain and spinal cord)
Present-case	F	14	Cough; fever; Chest pain. Some bouts of hemoptysis	Mass in the right middle lobe	Right middle lobe	Complete occlusion of the right middle lobe bronchus	Close funnel-shaped stenosis	Extension to pericardium, right atrium and pulmonary veins. No lymph nodes metastases	Explorative thoracotomy	–	10 cycles CYVADIC	Died 12 months after thoracotomy

M = male; F = female; ? = Not stated; – = Not performed

On bronchoscopic examination the right middle lobe bronchus was completely occluded and stenosed by a lipomatous vegetation which bled when touched with submucosal infiltration evident at the level of the right intermediate bronchus just before the origin of the right middle lobe bronchus.

Biopsy specimens contained only necrotic tissue so histologic diagnosis was not possible. The mass was then studied by bronchography and pulmonary angiography. On bronchography, a tight, funnel-shaped stenosis was observed in the distal part of the right intermediate bronchus with very scanty passage of the contrast media in the right middle lobe bronchus (Fig. 2 a, b). On pulmonary angiography (Fig. 2 c) there was constriction of third order vessels encased within the parenchymal lesion. Explorative thoracotomy showed a large right hilar tumour invading the pericardium, extending well into the right atrium and involving the pulmonary vessels.

Microscopic studies of the biopsy specimens showed neoplastic proliferation with spindle-shaped cells containing eosinophilic cytoplasm, organized separately or in parallel, in undulating whorls scattered between the normal smooth muscle fibres of the bronchial wall (Fig. 3, a, b).

There was no tumour spread to the resected lymph nodes. Final histologic diagnosis was that of a smooth muscle sarcoma (leiomyosarcoma).

Since extension of the tumour to the mediastinum made surgical resection or radiotherapy impossible, the patient underwent polychemotherapy (10 cycles of CYVADIC - DTIC + VCR + ADR + CPM - every 28 days) up to death 12 months after histologic diagnosis.

Discussion

Primary lung leiomyosarcomas are rare malignant tumours both in adults [1, 2, 7, 10] and children [2-6, 8, 9, 11]. The tumour arises from smooth muscle tissue present both in the bronchi and blood vessels; it generally spares lymphatics (no case of lymphatic extension has been reported in the pediatric age) and spreads by the blood-stream [7, 10] with secondary localisation in other organs including the heart [5], the other lung and brain [9]. We have found only nine pediatric patients, four males and five females, from a newborn baby to 11 years of age, described in the literature.

Clinical symptomatology in all reported cases, including our, is not clear cut at the onset, being characterized by cough, fever, wheezing, chest pain, increasing respiratory difficulty leading to dyspnea. In addition, nonspecific symptoms including weight-loss, at times recurrent pneumonia and hemoptysis have been observed. In all patients, an initial diagnosis of "neoplasm" has been made after chest X-ray which always showed the presence of a mass of varying size with well-defined borders, or atelectasis, associated with hyperventilation of the opposite lung in only one instance [11].

Bronchoscopy has usually revealed a mass obstructing the bronchial lumen, often with features of a chronic granulomatous infection. Biopsy per-

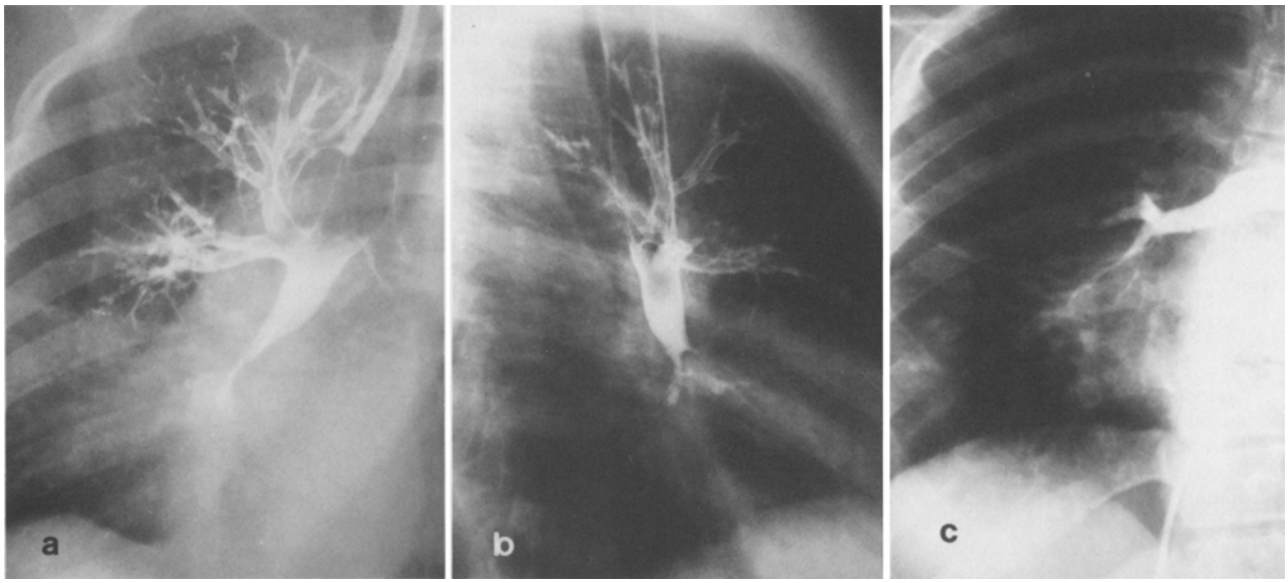

Fig. 2. a and b Funnel-shaped stenosis of the distal portion of the right intermediate bronchus. **c** Very thin and constricted third order vessels within the mass at angiography

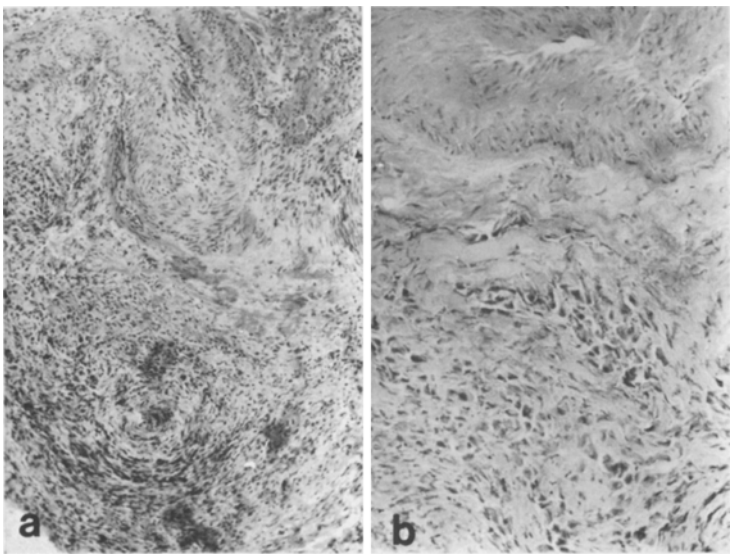

Fig. 3 a and b Microscopic specimens (**a** H. E. × 100, **b** × 350). Neoplastic proliferation of spindle-shaped cells organized separately or in undulating whorls amidst normal smooth fibers of bronchial wall

formed during bronchoscopy in only one case [4] allowed exact diagnosis (origin and type) of tumour in question. In all other cases, diagnosis of tumour type was made at surgery or autopsy. All cases reported in the literature have undergone surgical resection apart from one of the cases reported by Holinger [4], the newborn who died shortly after birth reported by Guccion [2] and the one described here.

In Holinger's case, extension of the tumour to the carina did not permit resection so the patient was treated with local radiotherapy but died of untreatable anemia a few months later. In the present case, extension of the tumour to the pericardium, right atrium and pulmonary vessels, rendered surgical resection or radiotherapy impossible, but polychemotherapy permitted survival for 12 months.

In all other cases (Table 1) results of surgery have been good: only two patients died, one of cardiac arrest during surgery [11] and one [9], who was among the youngest in the series, died 15 months after tumour resection, following a postoperative period complicated by metastases to the other lung (treated by radio-chemotherapy) and brain (right cerebellar hemisphere). Death was attributed to acute hydrocephalus secondary to posterior fossa surgery [9].

In this rare group of malignant tumours early di-

agnosis permits complete surgical resection of the mass and is associated with the best prognosis. Results of surgery might be further improved by addition of radio-and/or chemotherapy, at least in selected patients.

References

1. Dowell AR (1974) Primary pulmonary leiomyosarcoma: report of two cases and review of the literature. Ann Thorac Surg 17: 384
2. Guccion JG, Rosen SH (1972) Bronchopulmonary leiomyosarcoma and fibrosarcoma. A study of 32 cases and review of the literature. Cancer 30: 836
3. Hartman GE, Shochat SJ (1983) Primary pulmonary neoplasm of childhood: a review. Ann Thorac Surg 36: 108
4. Holinger PH, Slaughter DP, Novak FJ III (1950) Unusual tumors obstructing the lower respiratory tract of infants and children. Trans Am Acad Ophthalmol Otolaryngol 54: 223
5. Killingworth WP, McReynolds GS, Harrison AW (1953) Pulmonary leimyosarcoma in a child. J Pediatr 42: 466
6. Lawson EE, Goldstein IR (1971) Bronchial leiomyosarcoma in a child. J Pediatr Surg 6: 179
7. Morgan PGM, Ball J (1980) Pulmonary leiomyosarcomas. Br J Dis Chest 74: 245
8. Merritt JW, Parker KR (1957) Intrathoracic leiomyosarcoma. Can Med Ass J 77: 1031
9. Ownby D, Lyon G, Spock A (1976) Primary leiomyosarcoma of the lung in childhood. Am J Dis Child 130: 1132
10. Ramanathan T (1974) Primary leiomyosarcoma of the lung. Thorax 29: 482
11. Watson WL, Anlyan AJ (1954) Primary leiomyosarcoma of the lung. Cancer 7: 250

Date of acceptance: 14 July 1985

Dr. G. Beluffi
Department of Radiodiagnosis
Section of Pediatric Radiodiagnosis
Pediatric Clinics of the University
IRCCS Policlinico "S. Matteo"
I-27100 Pavia PV
Italy

Literature in pediatric radiology (continued from p. 237)

Pädiatrische Praxis (München)

Was leistet die kraniale Computertomographie bei der Diagnostik von Verhaltensauffälligkeiten im Kindes- und Jugendalter? Lehmkuhl, G. et al. (Kinder- und Jugendpsychiatrie, Klin. des Zentralinst. f. Seelische Gesundheit, Postfach 5970, Mannheim 1, FRG) **31,** 621 (1985)

Hüftsonographie und Vorsorgeuntersuchung. Enders, E. (Kinderklin. des Kreiskrankenhauses, Seilerweg 29, D-6430 Bad Hersfeld, FRG) **31,** 699 (1985)

Stellenwert der Sonographie bei der Differentialdiagnose der Hepatomegalie. Deeg, K.H., Zeilinger, G. (Univ.-Kinderklin., Loschgestr. 15, D-8520 Erlangen, FRG) **31,** 707 (1985)

Die Pericarditis constrictiva im Kindesalter. Brode, P. et al. (Kardiol. Abt. der Johanniter-Kinderklin., Arnold-Janssen-Str. 29, D-5205 St. Augustin 1, FRG) **32,** 19 (1985)

Streßfrakturen im Kindesalter. Schneider, K. et al. (Univ.-Kinderklin. München, Lindwurmstr. 4, D-8000 München 2, FRG) **32,** 139 (1985)

Ventrale Atlasdislokation (atlanto-axiale Instabilität) bei Kindern mit einer Trisomie 21 (M. Down). Storm, W. (Kinderklin. des St. Vincenz-Krankenhauses, Am Busdorf 4a, D-4790 Paderborn, FRG) **32,** 149 (1985)

Akutes Abdomen im Kindesalter. Diagnostische Maßnahmen und Operationsindikation. Willital, G. H. (Klinik und Poliklinik für Kinder- und Neugeborenenchirurgie, Univ. Münster, Albert-Schweitzer-Str. 33, D-4400 Münster, FRG) **32,** 455 (1985)

Odontogenes Fibrom der Mandibula bei einem 2 Jahre alten Kind. Kees, A. et al. (Chirurg. Abtlg. d. Mautner Markhofschen Kinderspit., Baumgasse 75, A-1030 Wien, Austria) **32,** 479 (1985)

Formen und Bedeutung von Trachealstenosen. Geißler, W. et al. (Chirurg. Abtlg. des Mautner-Markhofschen Kinderspit., Baumgasse 75, A-1030 Wien, Austria) **32,** 487 (1985)

Pertussis-Enzephalopathie. Zur Wertigkeit von Computertomogramm und klinischem Befund. Kosenow, W. (Kinderklinik der Städt. Krankenanstalten, Lutherplatz 40, D-4150 Krefeld, FRG) **32,** 525 (1985)

Radiologe (Berlin)

Heutiger Stellenwert und Grenzen der Sonographie in der Diagnostik der hypertrophen Pylorusstenose. Langer, R., Kaufmann, H.J. (Abt. f. Päd.-Rad., Klinikum Charlottenburg der Freien Univ. Berlin, Heubnerweg 6, D-1000 Berlin 19, FRG) **25,** 396 (1985)

Radiologie (Berlin)

Bronchial syndrome in infants. Fauré, Cl. (Hôp. Trousseau, Serv. de Rad., 8 à 28, ave. du Dr. A. Netter, F-75571 Paris, France) **5,** 135 (1985)

Chest developmental pathology in pediatric age. Bellini, F., Cheli, E. (Serv. de Rad., Clin. Ped., Univ. de Milan, Via Commenda 9, I - 10122 Milan, Italy) **5,** 175 (1985)

Apport de l'exploration tomodensitométrique du thorax en pneumologie pédiatrique. Joffre, F. et al. (Serv. de Rad., CHU Rangueil, Chemin du Vallon, F-31054 Toulouse, France) **5,** 209 (1985)

Leistungsfähigkeit und Auswirkungen der neuroradiologischen Diagnostik bei cerebralen Erkrankungen im Kindesalter. Ludwig, B. (Abt. f. Neurorad., Klinikum der Johannes Gutenberg-Univ., Langenbeckstr. 1, D-6500 Mainz, FRG) **25,** 502 (1985)

Radiologische Diagnostik der intrakraniellen Aneurysmen und arteriovenösen Mißbildungen eines pädiatrischen Krankengutes. Thun, F., Schauseil-Zipf, U. (Rad.-Inst. und Poliklin. der Univ., Josef-Stelzmann-Str. 9, Haus 12/30, D-5000 Köln 41, FRG) **25,** 508 (1985)

Prämedikation und Risiken bei Computertomographien im Neugeborenen- und Säuglingsalter. Abel, M. (Univ.-Kinderklinik, Mathildenstr. 1, D-7800 Freiburg i. Br., FRG) **25,** 599 (1985)

RÖFO Fortschritte auf dem Gebiete der Röntgenstrahlen und der Nuklearmedizin (Stuttgart)

Die Bedeutung der Sonographie als Akutdiagnostikum und zur Verlaufsbeobachtung beim stumpfen Bauchtrauma im Kindesalter. Hager, J., Egender, G. (Abt. f. Kinderchir., I. Univ.-Klin. f. Chir., Anichstr. 35, A-6020 Innsbruck, Austria) **143,** 44 (1985)

Vergleich röntgenologischer und histologischer Befunde bei Osteosarkomen nach präoperativer Chemotherapie. Sommer, H.-J. et al. (Delling, G., Pathol. Inst. der Univ., Martinistr. 52, D-2000 Hamburg 20, FRG) **143,** 74 (1985)

Computed tomography of pulmonary blastoma in a child. Jaspers, M.J.J.R. et al. Bloem, J.L., Dept. of Diagn. Rad., Univ. Hosp. Leiden, Rijnburgerweg 10, NL-2333 AA Leiden, Netherlands) **143,** 104 (1985)

Computertomographische Diagnose eines kongenitalen Optikuskoloboms. Irnberger, Th. (Röntgendiagn. Zentralinst. der Landeskrankenanstalten Salzburg, Müllnerhaupstr. 48, A-5020 Salzburg, Austria) **143,** 112 (1985)

(continued on p. 247)