The Radiological Appearance and Incidence of Lymphoma*

B. J. CREMIN, M.B., B.S., D.R.O.G., M.C.R.A., F.F.R., *Principal Specialist in Radiology, Red Cross War Memorial Children's Hospital and University of Cape Town*

SUMMARY

No short account of lymphomas will satisfy all physicians or cover their protean radiological manifestations. Of the lymphomas, Hodgkin's disease and lymphosarcoma present with the most common and consistent features. The appearances and frequency at their common sites is reviewed.

S. Afr. Med. J., **45**, 1360 (1971).

The radiological appearances and incidence of Hodgkin's disease† and lymphosarcomas as they affect lymph nodes and lymphatic tissues in the chest, abdomen and bones are presented and reviewed. Neither the lymphangiogram appearances nor the rare involvement of the intrinsic renal tract or central nervous system will be discussed.

The maximum frequency of Hodgkin's disease as judged by the age of onset is the third decade (20 - 29 years) but the mean age is the next decade (31 - 39 years).[1] Hodgkin's disease is uncommon in the first decade of life but a description in children will be given where the appearance or involvement differs from that in adults.

CHEST

The radiological appearances can be conveniently divided into mediastinal gland enlargement and parenchymal lesions.

Mediastinal Gland Involvement

The incidence of mediastinal gland involvement in Hodgkin's disease varies from 56% - 63%.[2,3] In their analysis of 154 proven cases from the Middlesex Hospital, Fisher *et al.*[2] reported that 56% (85 cases) of patients had enlarged mediastinal glands at some time in their illness. In the great majority of these patients glandular enlargement preceded symptoms by months or years. Just over one-third of their cases had glandular enlargement in their first 3 months, one-third within 2 years and in just under one-third, the glands became visible within the next 4 years.

In just over one-third of cases, the glandular involvement was in root glands (Fig. 1). This may be asymmetrical or unilateral and is a differentiating feature from sarcoid. In one-third of cases, the enlargement was in paratracheal glands (Fig. 2). These frequently have a lobulated or bossalated appearance and are usually the first glands to be involved.[2,4] One-fifth of cases had anterior mediastinal gland involvement, and this may mimic thymic tumours. The remaining 13% of cases had posterior mediastinal gland involvement, which may need penetrated or oblique views to demonstrate them although a barium swallow showing indentation of the oesophagus, may be helpful. The glands do not calcify unless there has been radiotherapy. Any of the reticuloses or leukaemia, particularly chronic lymphatic leukaemia, may give similar glandular enlargement to Hodgkin's disease but in these conditions parenchymal lesions are less common.[5] The diagnosis of Hodgkin's disease should always be established by biopsy as hilar and mediastinal lymph gland enlargement may be caused by primary or secondary neoplasms of many kinds as well as by other forms of lymphoma, tuberculosis and sarcoidosis.[6]

Parenchymal Involvement

The pulmonary appearances are pleomorphic and the pattern may change either spontaneously or following therapy.[5] Parenchymal involvement does not form a presenting feature but it occurs in one-third of all cases

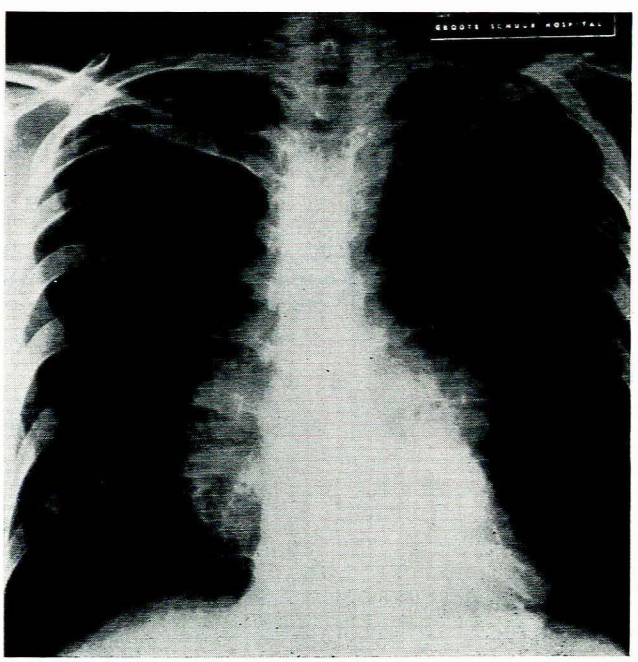

Fig. 1. Enlarged paratracheal and mediastinal glands. This has about a 60% occurrence and is the commonest manifestation of Hodgkin's disease.

*Date received: 24 August 1971.

†'Thomas' Hodgkin: his life and times' by the same author will appear shortly in the *S.A.M.J.* in the History of Medicine series.

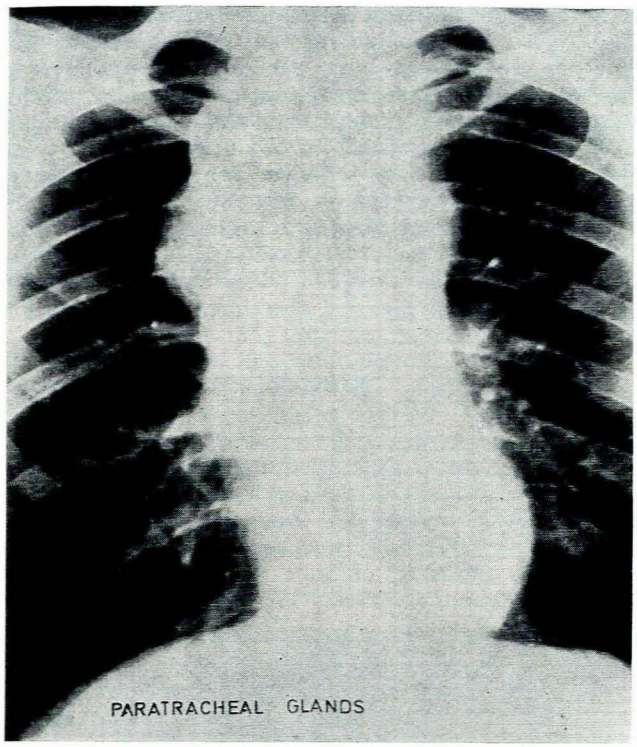

Fig. 2. Paratracheal glands with typical bosselated appearance. This may be an early and even an asymptomatic appearance.

and 60% of these cases are involved within 2 years of diagnosis. A simultaneous involvement of mediastinal glands occurs and early symptoms are cough, dyspnoea and malaise. The lesions may be briefly described as miliary, nodular, lobar and lymphangitic but a better account of the appearances and incidence may be given by adapting the classification used by Fisher *et al.*[2]

The parenchymal lesions may present as:

Shadows linear from root glands—85% of cases (Fig. 3). This is by far the commonest appearance and consists of continuous linear streaking which may closely resemble lymphangitis carcinomatosa. In lymphomas at this stage the glandular enlargement is usually more prominent than in carcinoma.

Shadows separate from root glands—'discontinuous lesions' (Fig. 4) 10% of cases. These lesions have apparently normal lung tissue between them and the root glands. The lesions may be isolated or form small groups. When rounded they may simulate secondary deposits but a blurring of the edges, possibly from interstitial infiltration, is often a distinguishing feature. The appearances may be very variable and a diffuse pattern may simulate bronchopneumonia or a coarse miliary stippling may resemble tuberculosis or sarcoidosis.

Bronchial glands—'sausage lesions', 5% of cases (Fig. 5). These irregular sausage like masses are formed from enlargement of nodes and lymphoid tissue around the bronchi and are seen as elongated shadows around the main bronchi.

Massive—segmental or lobar involvement may occur from a coalescence of smaller areas or a direct invasion from the mediastinum. These features are usually terminal. The masses may cavitate forming a lesion with thick-walled ragged interior (Fig. 6), or they may resemble a carcinoma. Tomography will show that the bronchi tend to remain patent and this is a helpful differentiating feature.

Pleural involvement is not uncommon and has a 28% incidence.[7] The pleurae have a rich lymphatic supply but in localized effusions without parenchymal lesions the adjacent ribs should be carefully scrutinized. Pericardiac involvement may occur from direct invasion from mediastinal glands and this event may be terminal. It is as well to remember that it may occur as a secondary infection in children who are having steroid therapy. Diaphragmatic paralysis is uncommon and this is a distinguishing feature from carcinoma in which it is relatively more frequent.[7]

INTESTINAL TRACT

In Hodgkin's disease the gastro-intestinal tract is only rarely exclusively involved.[8] More frequently the gastro-

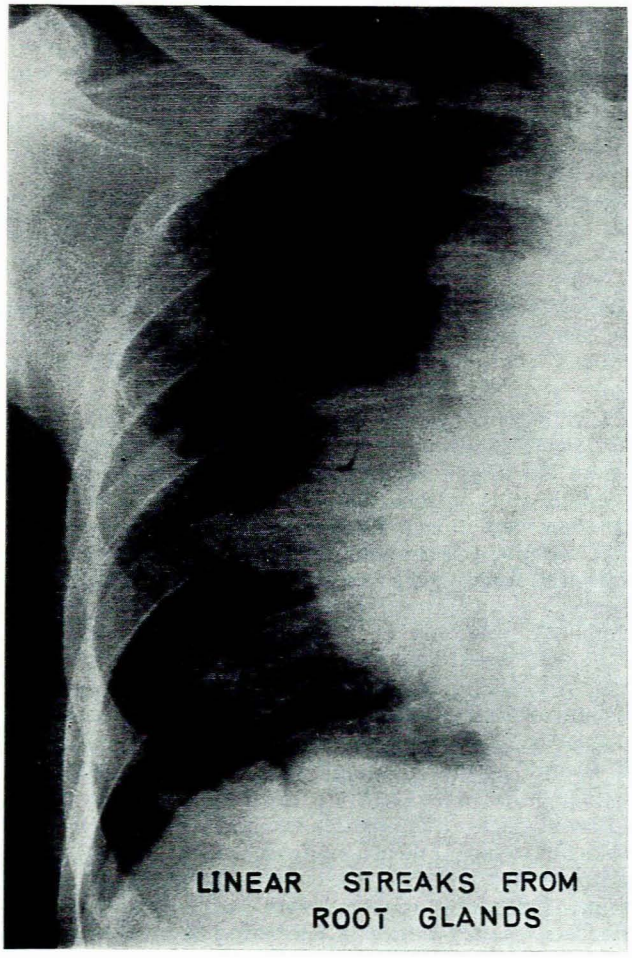

Fig. 3. Parenchymal involvement. Linear streaks radiating out from mediastinal root glands. This is the commonest appearance of lung tissue involvement.

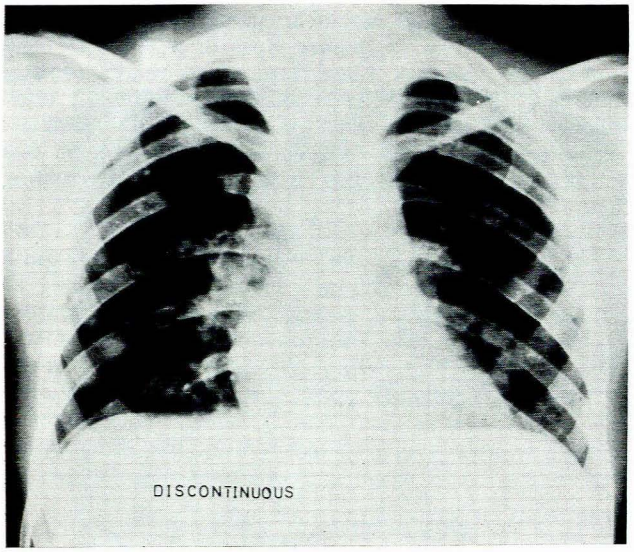

Fig. 4. Scattered 'discontinuous' lesions, that is, lesions separated from lung roots. Appearances are very variable.

intestinal symptoms are due to glandular enlargement or arise from systemic effects of the disease.[1] It is worth noting that 1 in every 5 patients with Hodgkin's disease has tumour deposits in the gastro-intestinal tract and that 1 in every 10 patients dies from gastro-intestinal involvement. The oesophagus and colon are rarely affected. The stomach is the most common site for isolated gastro-intestinal lesions. These comprise 2% of total gastric malignancy.[1] The lesions may be polypoid, ulcerating or infiltrating. On barium meal they may produce localized tumours, or appear as filling defects with smooth margins. Radiologically the differentiation from carcinoma may be impossible. The lesions involve predominantly the antrum but diffuse involvement giving a linitis plastica appearance may occur in 20% of cases.[8]

Lymphosarcoma may arise as a primary condition in the small intestine, particularly in the terminal ileum, where the largest aggregate of lymphoid tissue exists. In children it is a relatively frequent lesion in the small intestine, and it may simulate appendicitis, or may form the lead point in an intussusception. A good prognosis, following surgery and radical radiotherapy in cases limited to gut and mesenteric nodes has been reported.[18] Benign lymphoid polyposis is an unrelated condition due to localized hypertrophy of lymphoid follicles. It is usually seen in the colon and may give a soap bubble appearance to the mucosa, particularly on double contrast enema.[11]

A convenient descriptive classification of lymphosarcoma of the gut cannot be made. However, the macroscopical pathological appearances are often reflected in a barium follow-through examination. When the mesentery is diffusely involved, then a radiological and clinical pattern of sprue develops but the commonest lesion (60%) is a diffuse infiltration of the bowel wall.[8] This may cause irregular segmentation with narrowing or aneurysmal dilation from local infiltration of the bowel wall. The mucosal folds tend to be flattended, effaced or thickened. The polypoid masses may be discrete and intraluminal or may form multiple nodular defects to give a scalloped appearance to the mucosal pattern.[12] An endo-exocentric type involving the terminal ileum may form a tortuous tract through the narrowed lumen and may simulate Crohn's disease,[12] particularly if the latter lacks skip areas or saw tooth marginal ulceration.[10] Involvement of caecum may cause a mass in which differentiation from carcinoma, tuberculosis and amoebiasis may not be possible. In general terms Hodgkin's disease is less common than lymphosarcoma in the small bowel and it is more likely to be associated with systemic disease. It is also more likely to cause fibrosis and therefore obstruction from stenosis.[12]

BONES

In Hodgkin's disease lesions may appear in bones without any apparent mediastinal gland involvement.[13] Localized bone pain fequently precedes the X-ray appearances. The bony incidence in over 800 cases has been quoted as 14%[13] although this is likely to be an underestimation. At autopsy the incidence of bone involvement is at least 49%.[14] Bone lesions are usually considered a bad prognostic

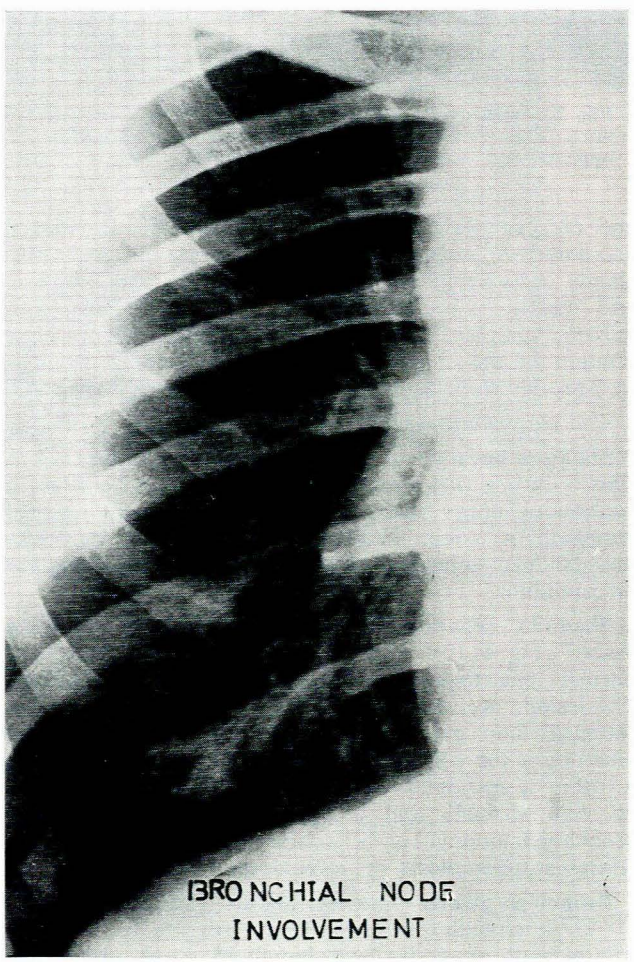

Fig. 5. 'Sausage-like' masses of lymphoid tissue that surround main bronchi and extend into lungs.

sign but paradoxically Stuhlbarg and Ellis[15] in a review of 179 cases found that those with bony lesions had a longer survival rate. It is likely that the bony involvement appears late and is thus commonest in patients with the longest survival. After their appearance half the patients die within 6 months.[16] The lesions are usually multiple and the commonest appearance is of a mixed osteoblastic and osteolytic change. In an assessment of 257 cases, Vieta *et al.*[14] gave the appearance figures as 57% mixed, 28% osteolytic and 14% osteoblastic. The sclerotic types do not enlarge bones but the lytic types, which usually involve the flat bones such as ileum, sternum and ribs may cause expansion. Radiotherapy may cause the appearances to revert to normal or cause sclerosis.[14] The axial skeleton is predominantly involved with the greatest incidence in lower dorsal or upper lumbar spine.[13] Associated paravertebral soft tissue masses may occur and the dorsal spine is involved in 50% of bone lesions.[2] The spine, sternum and sacro-iliac joints may become involved from adjacent glands, and this may cause a characteristic anterior notching of the involved vertebral body. The involvement of the ribs is also frequently the result of direct pleural spread and in these cases the rib may apparently vanish. The vertebral lesions are predominantly mixed or sclerotic,[13] and may cause collapse although this is not usually complete. In a series of 16 cases of single, sclerotic (ivory) vertebra,[7] the causes were found to be 8 focal Paget's disease, 6 lymphoma and 2 metastatic carcinoma. This incidence of Paget's disease will vary with geographical location. It may be differentiated by its coarser trabeculation, enlargement in the anterior posterior diameter and sometimes window framing from subcortical sclerosis. Involvement of the long bones occurs predominantly in the proximal marrow areas. A diffuse pattern of multiple small linear translucencies in lymphosarcomas may also occur in all forms of malignant lymphoma.[13] These 'rain drops' tend to run in the long axis of the shaft and may cause an interior scalloping of the cortex.

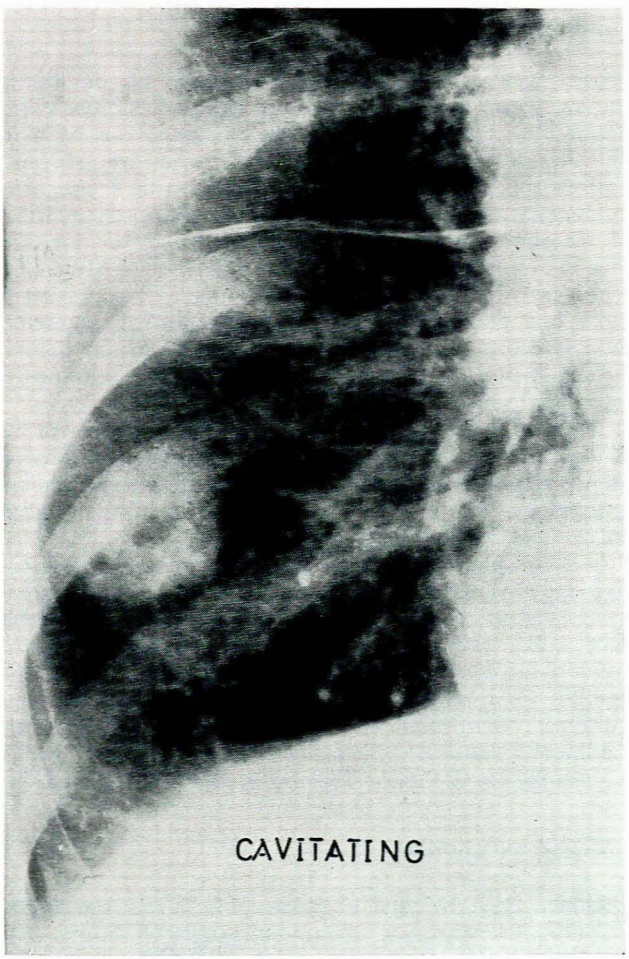

Fig. 6. Cavitating lesions, which may occur in small or large lesions and should arouse the suspicion of Hodgkin's disease.

The appearance of lymphosarcoma and diffuse reticular celled sarcoma is similar.[13] In general when bone is involved these cases have a poor prognosis. Typically they cause widespread lytic areas and may be responsible for pathological fractures. Lymphosarcoma has its maximum incidence in the age groups below and above those in which Hodgkin's predominates. In children there is an 8% transformation of lymphosarcoma into leukaemia.[13] Burkitt's lymphoma is a clinicopathological entity which has been reported in the facial bones of children from predominantly Central Africa and New Guinea.[19] Its description is beyond the scope of this article, but it may have an epidemiologic basis and is related to the Ebstein Barr virus and immunity to malaria[17] which would explain its geographic distribution.

Reticulum cell sarcomas may occur as a primary bone tumour and as such is beyond our scope except to state that it is a radio-sensitive tumour of variable growth and malignancy, which has a predilection for the long bones of adults.[13]

I would like to thank Drs B. Myers and S. Goldberg for their help.

REFERENCES

1. Wintrobe, M. M. (1967): *Clinical Hematology*. Philadelphia. Lea and Febiger.
2. Fisher, A. M. H., Kendell, B. and Van Leuven, B. D. (1962): Clin. Radiol., **13**, 115.
3. Wolpaw, S. E., Higley, C. S. and Hauser, H. (1944): Amer. J. Roentgenol., **52**, 374.
4. Stolberg, H. O., Patt, N. L., MacEwen, K. F., Warrick, O. H. and Brown, T. C. (1964): *Ibid.*, **92**, 96.
5. Sutton, D. (1969): *A Textbook of Radiology*, pp. 314 and 367. Edinburgh: E. & S. Livingstone.
6. Scadding, J. G. (1967): Proc. Roy. Soc. Med., **60**, 737.
7. Dennis, J. M. (1961): Radiology, **77**, 618.
8. Portmann, U. V., Dunne, E. F. and Hazard, J. B. (1954): Amer. J. Roentgenol., **72**, 772.
9. Cornes, J. S. (1967): Proc. Roy. Soc. Med., **60**, 732.
10. Cupps, R. E., Hodgson, J. R., Dockerty, M. B. and Adson, M. A. (1969): Radiology, **92**, 1355.
11. Cremin, B. J. (1970): Clin. Radiol., **21**, 195.
12. Marshak, R. H. and Lindner, A. E. (1970): *Radiology of the Small Intestine*, pp. 396 - 440. Philadelphia: W. B. Saunders.
13. Mosely, J. E. (1963): *Bone Changes in Hematologic Disorders*. New York: Grune & Stratton.
14. Vieta, J. O., Friendell, H. L. and Craver, L. F. (1942): Radiology, **39**, 1.
15. Stuhlbarg, J. and Ellis, F. W. (1965): Amer. J. Roentgenol., **93**, 568.
16. Hoskins, E. O. L. (1967): Proc. Roy. Soc. Med., **60**, 729.
17. Watson, C. (1971): Personal communication.
18. Burkitt, D. and O'Conor, G. T. (1961): Cancer (Philad.), **14**, 258.
19. Jenkin, R. D. T., Sonley, M. J., Stephens, C. A., Darte, J. M. M. and Peters, M. V. (1969): Radiology, **92**, 763.