Shimane J. Med. Sci. Vol. 5, pp. 180–186, 1981

Hodgkin's Disease in the Nose

(Hodgkin's disease/nose)

TADAO WAKUTANI, YASUAKI MIYAKUNI, and HISAO KITAMURA

Department of Oto-Rhino-Laryngology, Shimane Medical University, Izumo 693, Japan

(Received July 13, 1981)

We report herein a very rare case of Hodgkin's disease which developed in the right nasal region and followed a peculiar clinical course.

A 56 year old Japanese woman complained of right nasal obstruction and bleeding.

The lesion first showed evidence of nasal polyp and progressed slowly, but became aggravated and advanced rapidly immediately before admission.

The clinical staging on admission was established as stage IIEA.

The histologic type changed from one of mixed cellularity at onset of the treatment to the lymphocytic depletion type as seen at autopsy.

Malignant lymphoma that occurs in the head and neck region is usually a reticulum cell sarcoma, and most often involves Waldeyer's ring.

On the other hand, a malignant tumor that occurs in the nasal cavity and paranasal sinuses is often a carcinoma. Cases of malignant lymphoma are relatively rare. The occurrence of Hodgkin's disease, in this anatomical region has seldom been reported in the literature.

We treated a patient with a primary nasal lesion and who died bearing the lymphocytic depletion type.

We report the results of our treatment, observations and autopsy findings, with reference to the literature.

CASE REPORT

A 54-year Japanese old woman was admitted on February 28, 1980 with right nasal obstruction and bleeding.

In the fall of 1978, she had a feeling of obstruction in the right ear and was treated by an ENT specialist. Diagnosed as having secretory otitis media and chronic sinusitis, she was treated and the symptoms improved.

In the fall of 1979, she had a recurrence of a feeling of obstruction in the right ear and nasal obstruction. The same physician detected a nasal polyp.

She complained of a right nasal obstruction and nasal bleeding from the beginning of February, 1980, and again consulted the same physician. As a malignant tumor was suspected she was referred to our clinic.

She herself noticed a tumor mass on the right neck.

The family history was not remarkable, but in her own medical history she

had had a toxemia related pregnancy at age 25, blindness in the right eye of an unknown cause at 31, cardiac disease at age 50 and viral pneumomia at age 53.

Clinical Findings

General findings including the chest and abdomen showd no marked change. The axillary and inguinal lymph nodes were not palpable.

Anterior rhinoscopy revealed that the right nasal cavity was filled with a red, irregular, hard hemorrhagic tumor mass, part of which was covered with white coating.

This tumor mass had a base in the middle meatus. A white nasal polyp

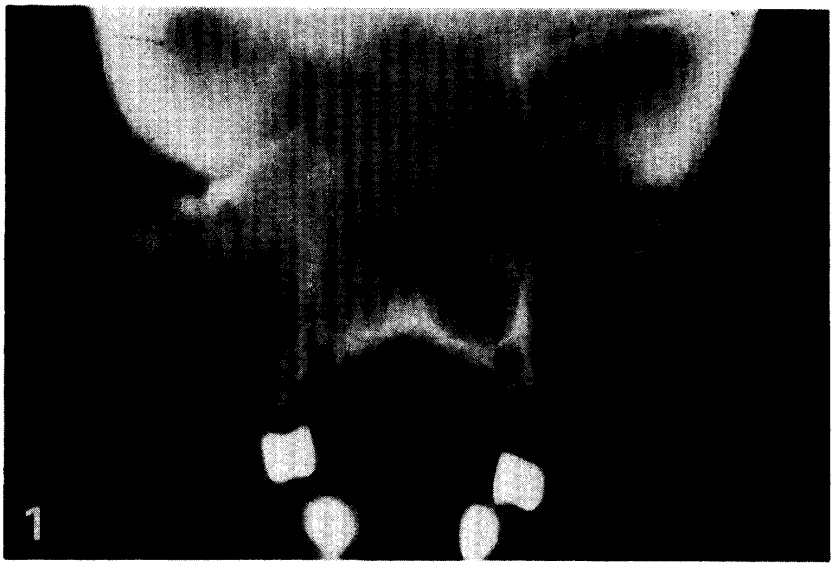

Fig. 1. Facial tomography shows a bony destruction of the medial wall in the right maxillary sinus.

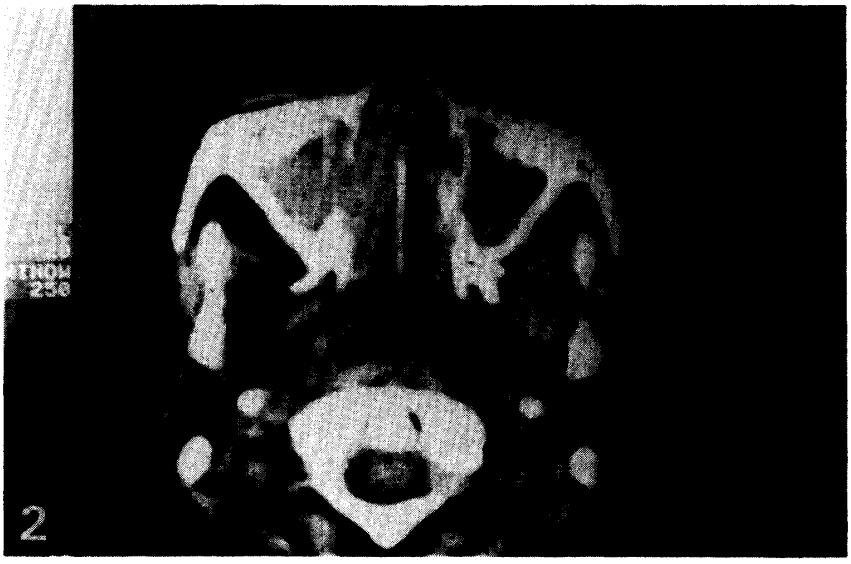

Fig. 2. CT scan shows a tumor mass lesion in the right nasal cavity and maxillary sinus.

was noted in the left middle meatus.

Pain and swelling in the cheek, alveolar process and hard palate, exophthalmos, and visual disturbance were not observed.

A dozen or so hard, enlarged, fixed nodules were palpable bilaterally, particularly in the right cervical region but there was no tenderness.

Tomographic examination of the nose revealed bone destruction on the medial wall of the right maxillary sinus and orbital floor. (Fig. 1) Similar findings were obtained on the CT.(Fig. 2)

Hematological examination, urinalysis and serum biochemistry were within normal limits. The serologic test for syphilis was negative.

Clinical Course

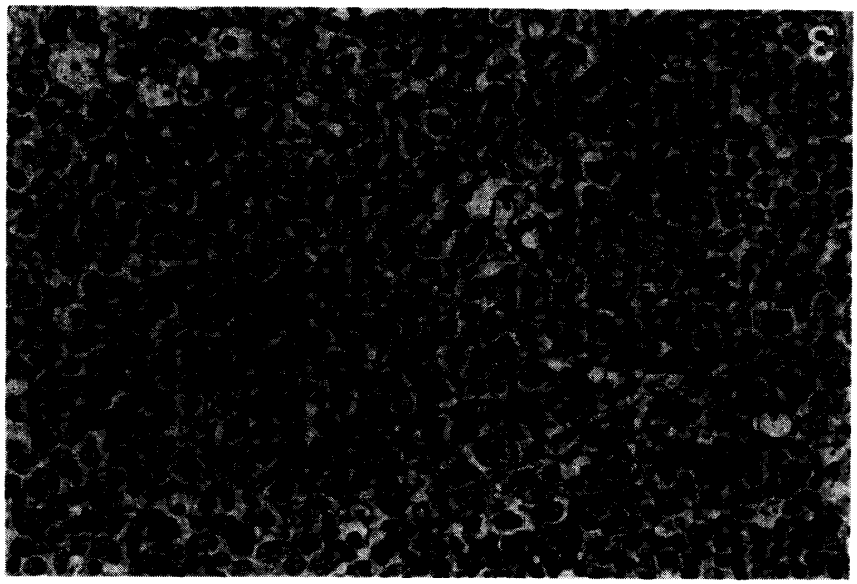

Fig. 3. A number of so-called Hodgkin's cells are seen in this specimen. (×200)

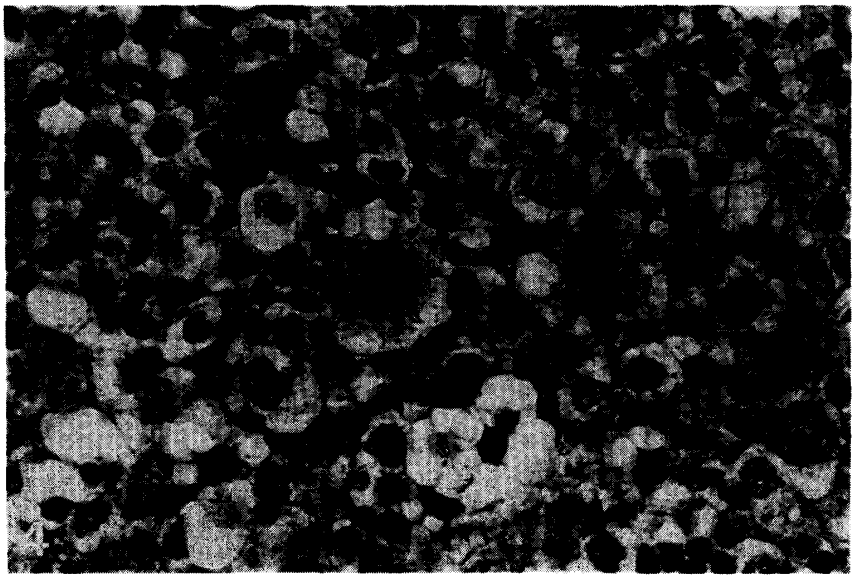

Fig. 4. The arrow indicates a Sternberg-Reed cell. (×400)

From the clinical findings above, the patient was suspected of having a malignant lymphoma. Biopsies of the tumor mass in the right nasal cavity and right cervical lymph node were performed. At the same time VER therapy (vincristine 1mg once a week, cyclophosphamide (Endoxan) 500mg once a week, betamethasone (Rinderon) 4mg twice a week) was initiated in accordance with our schedule (1).

Histological findings : Histological examination showed typical Hodgkin's disease (mixed cellularity type) with Sternberg-Reed cells co-existing with a number of so-called Hodgkin's cells. (Figs.3, 4)

Lymphangiography revealed no abnormal findings in the abdomen.

Accordingly, the clinical staging was established as stage IIEA.

External irradiation of ^{60}Co (200 R per day) to the right nasal cavity, paranasal sinus and bilateral cervical regions was started from March 10, 1980.

The patient responded well to radiation.

Following a total dose of 1200 R, the tumor mass in the nasal cavity had all but disappered, the left cervical lymph node was no longer palpable and only one lymph node was palpable in the right cervical region.

On Ga scintigraphy, however, a hot lesion was detected in the bilateral hilar regions, thus indicating hilar lymph node involvement.

External irradiation was also applied to that region.

The anti-tumor effect of the combined therapy was temporary and the involvement advanced rapidly to the bilateral inguinal regions, axillary nodes, skull etc.

In the meantime, a more potent chemotherapy was administered without effect, and she died on July 2. (Fig. 5)

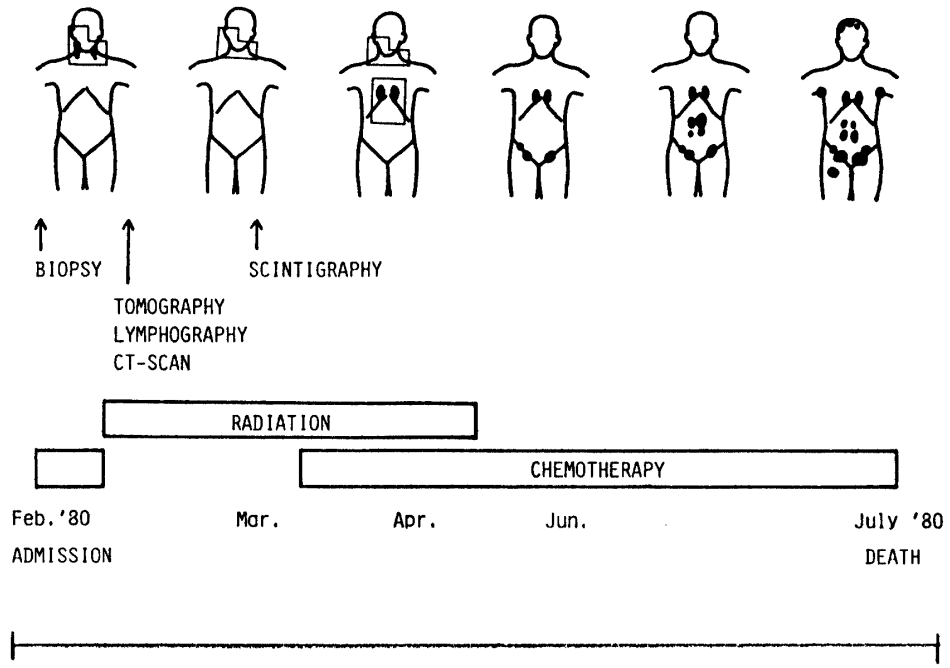

Fig. 5. Clinical course.

Autopsy findings :

1. Lymphocyte depletion type of Hodgkin's disease.
2. The involvement extended to (1) general lymph nodes, e. g., bilateral axillary, inguinal, hilar, peritracheal, iliac and para-aortic regions (2) both lungs (3) skull.
3. Of the irradiated organs, the nasal cavity and paranasal sinus were free of involvement.

DISCUSSION

Incidence

Hodgkin's disease may occur in any organ of the body. According to Tanimoto and Hattori (2), however, an increase in the incidence of non-Hodgkin's lymphoma, particularly reticulum cell sarcoma, among the cases of malignant lymphoma, is conspicuous and the incidence of Hodgkin's disease is leveling off and low in Japan compared with the occurrence in Europe and America.

Baroni and Malchiodi (3) reported that the primary lesion of Hodgkin's disease in Southern Europeans was in the cervical nodes followed by the supraclavicular nodes and axillary nodes.

In Japan, Amaki and Kitami (4) reported that the primary lesion was found in the cervical nodes, mediastinal structures, abdominal nodes and axillary nodes, in this order.

In the head and neck region, Hodgkin's disease occurs in the cervical nodes and Waldeyer's ring abound with lymphoid tissue.

According to Saito (5), Hodgkin's disease accounts for 10.4 percent (13 cases) in 124 cases of malignant lymphoma and by location an overwhelmingly large number (54%, 6 cases) is found in the cervical nodes.

Recently, Awataguchi (6) reported that of 27 cases of palatine tonsil, 24 were reticulum cell sarcoma and only two were Hodgkin's disease.

Hodgkin's disease seldom occurs in the nasal cavity and paranasal sinuses.

Stewart and Stuart (7) reported that there have been only seven such cases including theirs.

In Japan also, there have been no detailed reports except for two briefly introduced cases.

The case presented here is one of Hodgkin's disease in which the lesion at first showed findings of a nasal polyp and grew slowly, as was also found by others (7).

Clinical Staging

The classification used at present is one that was determined in 1971 (8).

Parameters include (1) detailed history, (2) detailed general findings, particularly findings in the lymphoid tissues, (3) biopsy of the primary lesion including lymph nodes, (4) chest x-ray including tomography, lymphangiography, Ga scintigraphy, computed tomography in addition to hematology,

urinalysis and serum biochemistry and (5) bone marrow examination and staging laparotomy.

Kaplan (9) maintained that lymphangiography, computed tomography and laparotomy with splenectomy are absolutely required to accurately determine the clinical staging.

Sterchi and Myers (10) stressed the necessity of staging laparatomy with multiple liver and lymph node biopsies and splenectomy.

The examinations should be done in parallel with the treatment, as in the present case, where it was not until Ga scintigraphy was performed that the hilar lesion was discovered.

In the present case, it was not clear whether the primary lesion was in the nasal cavity or in the paranasal sinus as the involvement occurred continuously through the soft tissue and thin bony plates.

Management

In studying basic therapeutic policies for Hodgkin's disease, it would be necessary to consider how this disease advances.

As a therapeutic policy for reticulum cell sarcoma, we treated this tumor as a general disease, even if it was of the localized type of stages I and II.

On the other hand, many researchers hold the view that Hodgkin's disease progresses in a monocentric manner.

However, Amaki and Kitami (4) maintained that there is the possibility of multicentric growth with the mixed cellularity and lymphocytic depletion type of Hodgkin's disease, as in the present case.

We are of the view that there are cases in which it is necessary to adopt a therapeutic policy with radiotherapy subordinated to chemotherapy, as in the case of reticulum cell sarcoma and depending on the histologic type.

Multidrug combination therapy is widely prescribed and a variety of combination therapies such as MOPP, COPP, VEMP, BEMP and BONP are available (11). When prescribing chemotherapy plus radiation, we use the VER therapy.

As to radiotherapy, a total dose of 3500—4000 R is applied to the field, including the adjacent lymphoid region for stages I and II, in Japan (4).

Kaplan (9) holds the same view as ours and recommends total lymphoid megavoltage radiotherapy or multidrug combination therapy, regardless of the stage.

Prognosis

Regarding the prognosis of this disease, Tanimoto and Hattori (2) showed that it is most closely related to spread of the staging of the lesion, that is, classification and mentioned the histologic type and possible presence of general symptoms as factors related to the prognosis.

With the histologic type, Watanabe *et. al.* (12) pointed out that in Japan, there are characteristically many cases of mixed cellularity and lymphocytic depletion types of which the prognosis is poor.

Colby and Warnke (13) compared initial and relapsed cases as to the histologic type and reported that the histologic type remained the same in most cases but changes to a form with a poor prognosis also occurred.

Such a change in the cellularity may be likened to the terminal stage of cancer in which cancer cells undergo a sarcomatous change.

The present case was of the stage IIEA type but changed to the histologic from the mixed cellularity type at the onset of the treatment then to the lymphocytic depletion type.

REFERENCES

1) Wakutani, T., Kobayashi, H., Kitamura, H., and Miyakuni, Y. (1981) Combined chemotherapy and radiotherapy for malignant lymphomas of the head and neck. *Shimane J. Med. Sci.* **5**, 69–76
2) Tanimoto, K. and Hattori, K. (1978) The epidemiology of malignant lymphomas in Japan. *Naika* **41**, 364–368 (in Japanese)
3) Baroni, C. D. and Malchiodi, F. (1980) Histology, age and sex distribution, and pathologic correlations of Hodgkin's disease. A study of 184 cases observed in Rome, Italy. *Cancer* **45**, 1549–1555
4) Amaki, J. and Kitami, Y. (1978) Hodgkin's disease. *Naika* **41**, 407–412 (in Japanese)
5) Saito, H. (1977) Some problems of the head and neck malignant lymphomas-Analysis of eighteen years of observation *Jibi Inkoka* **49**, 183–190 (in Japanese)
6) Awataguchi, S. (1979) Cancer of the airway. The Alumni Association of Department of Oto-Rhino-Laryngology, Hirosaki University School of Medicine, Hirosaki, Japan (in Japanese)
7) Stewart, I. A. and Stuart, A. E. (1971) Hodgkin's disease in the nose. *J. Laryngol. Otol.* **85**, 1069–1073
8) Carbone, P. P., Kaplan, H. S., Musshoff, K., Smithers, D. W., and Tubiana, M. (1971) Report of the committee on Hodgkin's disease staging. *Cancer Res.* **31**, 1860–1861
9) Kaplan, H. S. (1980) Hodgkin's disease : Unfolding concept concerning its nature, management and prognosis. *Cancer* **45**, 2439–2473
10) Sterchi, J. M. and Myers, R. T. (1980) Staging laparotomy in Hodgkin's disease. *Ann. Surg.* **191**, 570–575
11) Tanimoto, K. and Hattori, K. (1975) Current chemotherapeutic concept for malignant lymphomas. *Gan To Kagakuryoho* **2**, 753–761 (in Japanese)
12) Watanabe, S., Matsuda, M., Mikata, A., and Kageyama, K. (1979) Epidemiologic study for Hodgkin's disease. *Nippon Rinsho* **35**, 3673–3679 (in Japanese)
13) Colby, T. V. and Warnke, R. A. (1980) The histology of the initial relapse of Hodgkin's disease. *Cancer* **45**, 289–292