

Idiopathic Cervical Fibrosis—A New Member of IgG4-Related Sclerosing Diseases: Report of 4 Cases, 1 Complicated by Composite Lymphoma

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Abstract: Idiopathic cervical fibrosis is a rare tumefactive inflammatory-sclerosing lesion involving the soft tissues of the head and neck, and a proportion of patients also have synchronous or metachronous inflammatory fibrosclerosing lesions in other anatomic sites. The latter finding suggests that this entity may represent a member of IgG4-related sclerosing diseases. We report 4 cases to support this postulation. The patients were male adults aged 42 to 89 years, who presented with an infiltrative, firm cervical mass. Two patients also had IgG4-related chronic sclerosing sialadenitis of submandibular gland and lymphadenopathy. Histologically, the cervical soft tissue lesions had ill-defined borders, consisting of coalescent nodular lymphoid aggregates accompanied by a sclerotic stroma. Nerve infiltration, skeletal muscle invasion, and phlebitis were present. There was a significant increase in IgG4⁺ plasma cells (87 to 327 per high-power field, with IgG4/IgG ratio of 63% to 98%). In the soft tissue lesion of 1 patient, there were expansile foci comprising dense sheets of plasma cells and small lymphoid cells that exhibited κ light chain restriction and clonal immunoglobulin gene rearrangement, consistent with supervening extranodal marginal zone lymphoma. The adjacent lymph node from the same patient showed Epstein-Barr virus (EBV)-positive classical Hodgkin lymphoma with typical morphology and immunophenotype (CD30⁺, CD15⁺, PAX5⁺). Thus lymphoma can supervene in the chronic inflammatory background similar to that recently documented for IgG4-related sclerosing disease of the ocular adnexa.

Key Words: idiopathic cervical fibrosis, soft tissue, IgG4-related sclerosing disease, chronic sclerosing sialadenitis, extranodal marginal zone lymphoma, Hodgkin lymphoma, composite lymphoma

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Idiopathic cervical fibrosis, also known as sclerosing cervicitis and cervical fibrosclerosis, is a very rare mass-forming lesion involving the soft tissues of the head and neck, first described by Rice et al in 1975.^{8,22} The cause has been attributable in the past to infection, Hodgkin lymphoma, sarcoidosis, autoimmune disease, trauma or prior surgery, but most cases are considered idiopathic.⁷ It has also been suggested that the disease can represent a localized manifestation of a systemic condition,¹ because about 20% of patients also have other inflammatory fibrosclerosing lesions, such as sclerosing cholangitis, mediastinal fibrosis, retroperitoneal fibrosis, and orbital pseudotumor.^{13,14,20–22} Recently, most cases of the inflammatory fibrosclerosing lesions have been found to belong to the family of IgG4-related sclerosing diseases.^{3,9,26,27} We herein report 4 cases of idiopathic cervical fibrosis to confirm its relationship with IgG4-related sclerosing disease, with 1 patient complicated by a hitherto unreported phenomenon of supervening composite lymphoma.

MATERIALS AND METHODS

The 4 cases were retrieved from the consultation files of 2 of the authors (J.K.C.C. and W.C.) and surgical pathology archives of Queen Elizabeth Hospital, Hong Kong. Four-micron-thick sections were cut from formalin-fixed, paraffin-embedded blocks and stained with hematoxylin and eosin for light microscopic study. Immunohistochemical staining was done on the BOND-MAX-automated immunostainer (Vision Biosystems, Australia) using a polymer-based detection system. The following antibodies were applied: anti-IgG (1:250, IgGp, Novocastra), anti-IgG4 (1:500, HP6025, Zymed), CD20 (1:1,000, L26, Dako), anti-IgD (1:1000, antiserum, Dako), anti-IgA (1:1500, antiserum, Dako), anti-IgM (1:5000, antiserum, Dako), CD3 (1:150, PS1, Novocastra), CD21 (1:15, 2G9, Novocastra), CD10 (1:10, 56C6, Neomarkers), CD5 (1:50, 4C7, Novocastra), cyclin D1 (1:100, SP4, Neomarkers), BCL2 (1:500, Bcl-2-100, Zymed), CD30 (1:120, BerH2, Dako), CD15 (1:100, Carb-3, Dako), PAX-5 (1:25, 1EW, Novocastra), and immunoglobulin κ -light and λ -light chains (1:48,000, antiserum, Dako). In situ hybridization for Epstein-Barr

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virus (EBV) early RNA (EBER) was conducted on the BOND-MAX automated immunostainer using fluorescein-conjugated EBV probe (NCL-EBV, Novocastra).

For enumeration of IgG4⁺ and IgG⁺ cells, the areas with the highest density of positive cells were evaluated. Three high-power fields (HPF) in each histologic section were counted, and an average number of positive cells per HPF was calculated. One HPF covered an area of 0.196 mm² (Nikon microscope, ×40 objective, ×10 eyepiece, 20-mm field of view). An absolute IgG4⁺ cell count of more than 50 cells per HPF, together with a ratio of IgG4⁺/IgG⁺ cells above 40%, was considered immunophenotypic compatibility with IgG4-related sclerosing disease.

Polymerase chain reaction (PCR) was conducted in case 3 to look for clonal immunoglobulin gene rearrangements using Bio-Med 2 primers (Invivoscribe Technolo-

gies). DNA was extracted from selected areas in the paraffin blocks based on assessment of corresponding stained sections, from areas rich in Reed-Sternberg cells and B-cell lymphoma, respectively.

RESULTS

Clinical Features

All patients were males aged 42 to 89 years (mean 65.5 y). The clinical features are summarized in Table 1. The patients presented with neck masses, often clinically thought to be enlarged lymph nodes with suspicious features because of the ill-defined borders of the lesions. Biopsy or excision was performed, and 2 cases showed dramatic response of the residual mass lesions to steroid therapy (cases 2 and 3).

TABLE 1. Clinicopathologic Features of Patients With IgG4-related Cervical Fibrosis

Case	Sex/Age	Presentation	Treatment and Outcome	Blood Tests	IgG4 ⁺ /IgG ⁺ Cell Counts per HPF
1	M/42	Presented with 6-mo history of bilateral neck swellings. Examination showed multiple hard nodules in deep neck spaces of submandibular and submental regions, as confirmed on ultrasonography. The right submental mass was excised because of clinical suspicion of malignancy despite a negative fine needle aspiration cytology	Excision of right submental mass. Then lost to follow-up	Not available	Cervical soft tissue: 168/215 (78%)
2	M/60	Had history of asthma. Presented with left cervical mass. Ultrasound showed multiple small lymph nodes (most less than 1 cm) surrounded by thick layers of abnormal tissue. Ultrasound of abdomen and flexible laryngoscopy showed no abnormalities	Biopsy of left neck mass, followed by a short course of oral prednisolone, which resulted in marked reduction in the size of the neck mass. Well at 1 y	IgG4 (1 week after biopsy) 125 mg/dL	Cervical soft tissue: 87/138 (63%) Lymph node: 172/365 (47%)
3	M/89	History of chronic sclerosing sialadenitis of left submandibular gland (excised 5-y ago), left parotid mass (1 cm, not biopsied) and left orbital mass (1.5 cm, not biopsied). Presented with a 2.4-cm hard mass over the right lower neck and a 5-cm right submandibular mass. Imaging showed diffuse lymphadenopathy in cervical, mediastinal, axillary and paraaortic regions.	Excision of the right cervical and submandibular masses. Given steroid for 5 d, with prompt subsidence of all lymph nodes. Curative chemotherapy and radiotherapy refused owing to advanced age. Alive and well at 1 y	IgG4 (postoperation) 795 mg/dL IgG (preoperation) 2350 mg/dL Erythrocyte sedimentation rate 120 mm/h (< 32 mm/h)	Left submandibular gland: 340/347 (98%) Right submandibular gland: 296/348 (85%) Cervical soft tissue: 310/400 (78%) Right submandibular lymph node: 359/441 (81%)
4	M/71	History of chronic sclerosing sialadenitis of right submandibular gland and enlarged right submandibular lymph node (excised 1 y ago). Presented with nontender right upper cervical mass (3 cm) associated with adjacent lymph node enlargement (0.3 to 1.5 cm)	Excision of right upper cervical mass and lymph nodes. Recurrence of right upper cervical mass at 1 y, and the lesion (about 2 cm) remained static in size on follow-up at 5 y	Not available	Right submandibular lymph node: 251/255 (98%) Right submandibular gland: 233/265 (88%) Cervical soft tissue: 327/333 (98%) Cervical lymph node: 140/222 (63%)

Normal range: serum IgG4 0-135 mg/dL, IgG 700-1600 mg/dL.

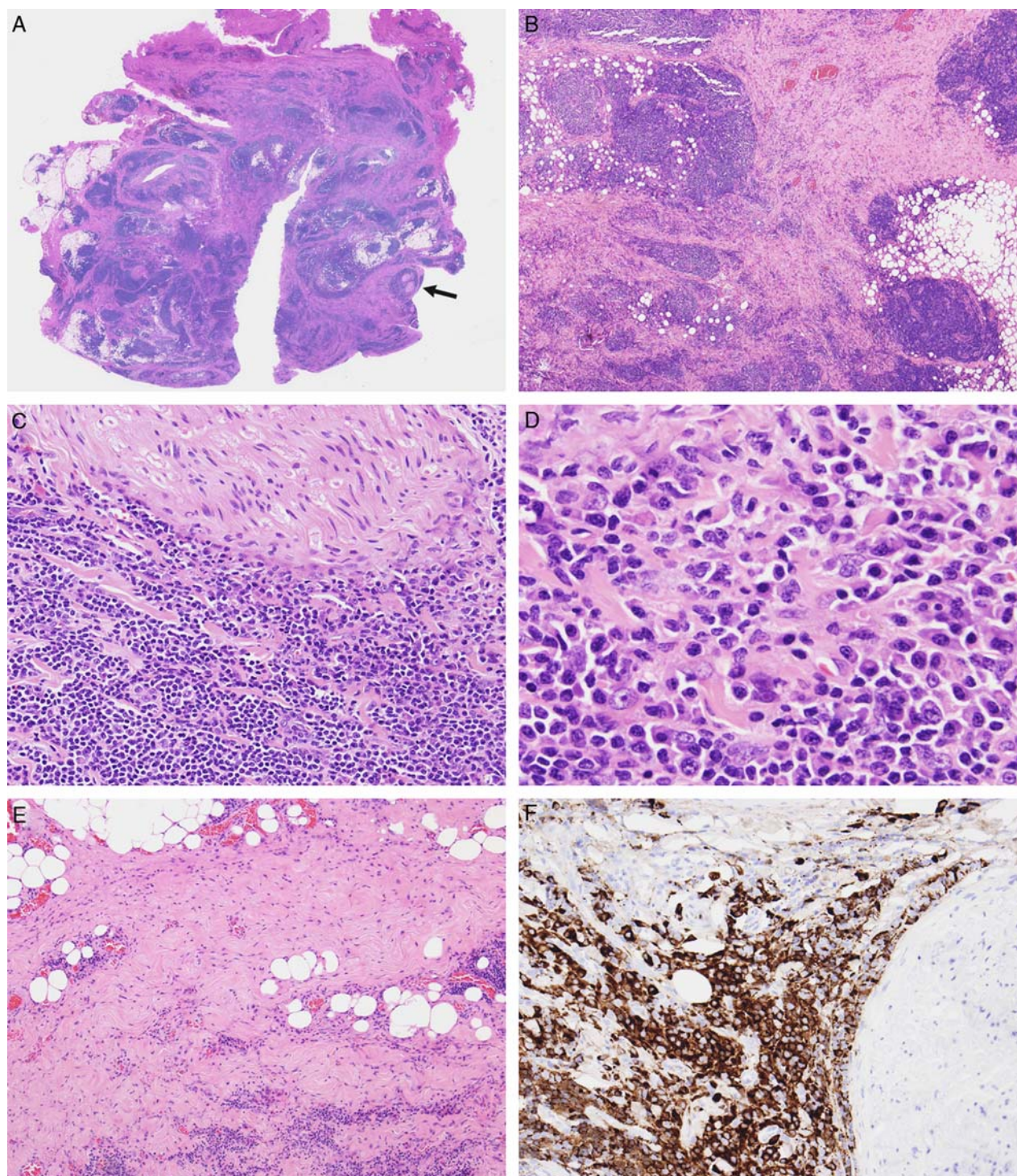


FIGURE 1. Soft tissue of case 2. A, Scanning view shows an infiltrative lesion comprising lymphoid aggregates and sclerotic stroma, with formation of dense lymphoid cuff around a nerve (arrow). B, Variable-sized lymphoid aggregates lie in a sclerotic stroma. C, Lymphoid cells encase a nerve (upper field). D, The lymphoid infiltrate comprises predominantly small lymphocytes and plasma cells. E, Some areas are formed predominantly by sclerotic tissue. F, Immunostaining for IgG4 shows many positive cells; this focus is adjacent to a nerve (right field).

Pathologic Findings

Cervical Soft Tissues (Cases 1, 2, 3, and 4)

The soft tissue masses had ill-defined borders, consisting of coalescent nodular lymphoid aggregates lying in a sclerotic stroma (Figs. 1–3). The former comprised reactive lymphoid follicles, small lymphocytes, and plasma cells, admixed with variable numbers of eosinophils. The lymphoplasmacytic infiltrates entrapped nerve bundles and formed dense cuffs around them (Figs. 1A, C, F, Fig. 3D). There was skeletal muscle invasion (Figs. 2A, B). Phlebitis with or without luminal obliteration was identified in all cases (Fig. 2D). The sclerotic stroma was hypocellular and composed of bands or thick trabeculae of collagen with few fibroblasts and myofibroblasts (Fig. 1E).

Immunostaining showed many CD3⁺ T cells in the inflammatory infiltrates, whereas CD20⁺ B cells were largely confined to the follicles. The plasma cells were

polytypic. IgG4⁺ plasma cells numbered 87 to 327 per HPF, constituting 63% to 98% of all IgG⁺ cells in the 4 cases (Fig. 1F, Fig. 3B) (Table 1).

In case 3, there were several expansile foci comprising diffuse sheets of mature plasma cells mixed with small lymphocytes and occasional large lymphoid cells (Fig. 3). The lymphoid cells (but not the plasma cells) were positive for CD20, and negative for CD3, CD5, BCL6, CD10, and cyclin D1. The plasma cells exhibited κ light chain restriction (Figs. 3E, F). IgG was expressed, but not IgA, IgM, or IgD; it is interesting to note that, 1 large aggregate of plasma cells that surrounded a nerve showed IgG4 expression (together with κ -light chain restriction). In situ hybridization for EBER was negative. Clonal immunoglobulin gene rearrangements were demonstrated by PCR. These foci were interpreted as extranodal marginal zone lymphoma of mucosa-associated lymphoid tissue, arising in a background of IgG4-related sclerosing disease.

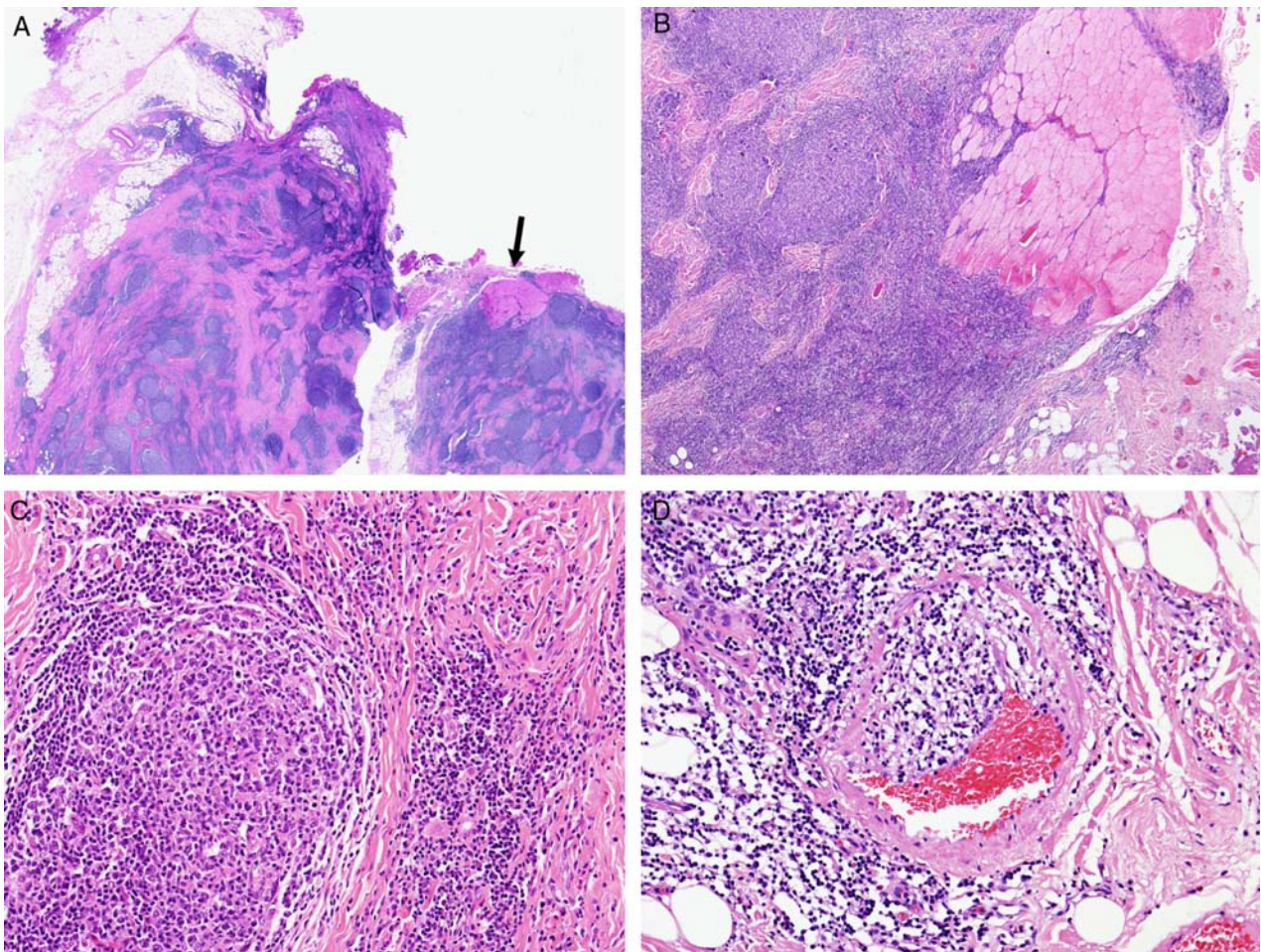


FIGURE 2. Soft tissue of case 1. A, The lesion, which comprises lymphoid aggregates in a sclerotic stroma, infiltrates into the adipose tissue. It also infiltrates skeletal muscle (arrow). B, The dense lymphoid infiltrate extends into and replaces skeletal muscle fibers. C, Lymphoid follicles occur in a sclerotic stroma. D, A vein shows subintimal and mural infiltration by lymphocytes and plasma cells. [full color online](#)

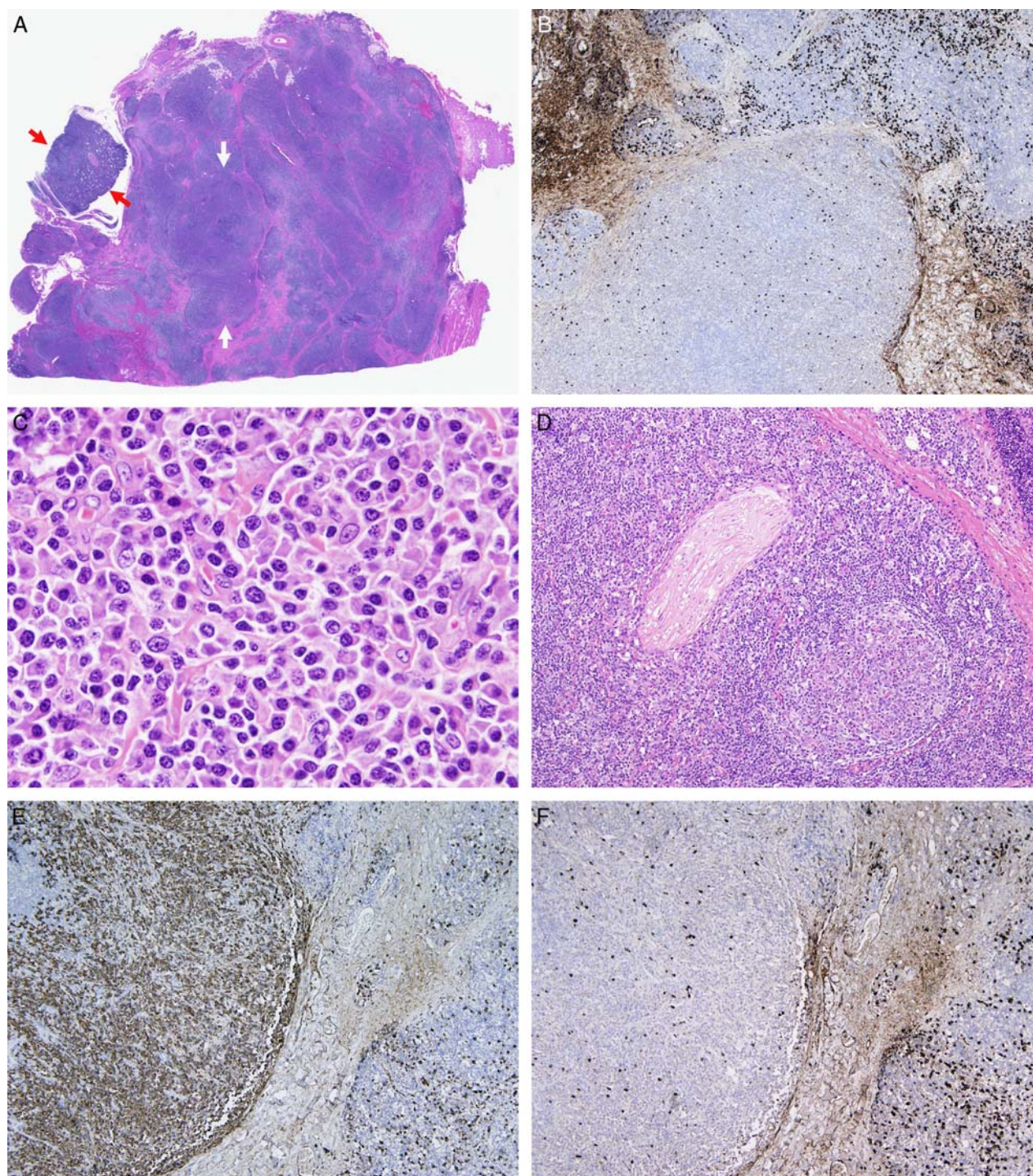


FIGURE 3. Soft tissue of case 3. A, Scanning view shows a lesion with poorly defined borders, comprising coalescent lymphoid aggregates in a sclerotic stroma. An expansile focus (white arrows) represents supervening lymphoma. Another focus (dark-colored arrows) also represents lymphoma, which entraps nerve. B, Immunostaining for IgG4 shows many positive cells in the background (upper and right field), but few positive cells in the expansile focus. C, The focus comprises a dense infiltrate of plasma cells, small lymphoid cells, and rare large cells. D, The background lesion comprises a mixed lymphoid infiltrate with scattered reactive lymphoid follicles. An entrapped nerve is seen. E and F, Immunostaining for κ and λ light chains, respectively. The expansile focus (left field) shows κ light chain restriction, while the background (right field) shows a polytypic mixture of κ -positive and λ -positive cells.

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Lymph Node (Case 2, 3, and 4)

In Cases 2 and 4, small lymph nodes embedded in or attached to the soft tissue specimen showed reactive follicular hyperplasia and moderate numbers of plasma cells in the interfollicular areas. The number of IgG4⁺ plasma cells ranged from 140 to 251 per HPF, constituting 47% to 98% of IgG⁺ cells. Features were compatible with type II IgG4-related lymphadenopathy.⁵

In Case 3, a lymph node measuring 18 × 11 mm was attached to the right submandibular gland. Approximately, 60% of nodal area showed effaced architecture, with a mottled appearance created by large cells with irregular nuclei, vesicular chromatin, and inclusion-like nucleoli scattered singly in a background of small lymphocytes, plasma cells, eosinophils, and histiocytes (Fig. 4). The large cells expressed CD30, CD15, and PAX-5 (weak), but not CD20, CD3, and immunoglobulins (Figs. 4E, F). These cells were positive for EBER (Fig. 4G). There were few admixed IgG4⁺ cells in such areas (Fig. 4D). No clonal rearrangements of immunoglobulin genes were found on PCR. The features were consistent with classical Hodgkin lymphoma, mixed cellularity type. In the preserved areas of the lymph node, numerous polytypic plasma cells were present between reactive follicles. There were 359 IgG4⁺ cells per HPF, constituting 81% of all IgG⁺ cells, compatible with type II IgG4-related lymphadenopathy (Fig. 4D).⁵

Submandibular Glands (Case 3 and 4)

The submandibular glands showed identical morphologic features. The lobular architecture was accentuated by thick fibrous septa. There was moderate lymphoplasmacytic infiltrate with a scattering of reactive lymphoid follicles in the parenchyma, accompanied by loss of acini and periductal sclerosis. No lymphoepithelial lesion was found. Immunostaining for CD3 and CD20 showed mixed populations of T and B cells. The B cells did not show diffuse sheets, aberrant immunophenotype or light chain restriction. The IgG4⁺ cell count was from 233 to 340 per HPF, with IgG4/IgG ratio of 85% to 98%, compatible with IgG4-related chronic sclerosing sialadenitis.

DISCUSSION

Idiopathic cervical fibrosis represents one form of tumefactive fibroinflammatory lesions of the head and neck, an entity most frequently affecting the sinonasal tract and parotid gland besides the cervical soft tissues, but also sometimes the mandible, tongue, face, cheek, and infratemporal fossa.^{8,21} The age at presentation is most commonly in the fifth to seventh decades with a predilection for male. The presenting symptoms, apart from a palpable mass, depend on the site of involvement. Neurologic symptoms, such as hoarseness, numbness, and shoulder drop are not uncommon owing to frequent direct nerve involvement.^{18,20,22} Local invasion of the meninges, skull base, orbit, and brain can also occur.¹¹ A favorable response to steroid therapy has been

documented.^{12,14} Some patients have synchronous or metachronous idiopathic hypertrophic pachymeningitis, orbital pseudotumor, mediastinal fibrosis, retroperitoneal fibrosis, or sclerosing cholangitis.^{10,13,14,19–22,24} These inflammatory fibrosclerosing lesions have in recent years been shown to represent members of IgG4-related sclerosing diseases.^{2,3,16,28} Thus the clinical findings strongly suggest that idiopathic cervical fibrosis belongs to this family of lesions.

In this series, the 4 cases of tumefactive lesions centered in the cervical soft tissues exhibit morphologic features compatible with idiopathic cervical fibrosis, but also characteristic of inflammatory fibrosclerosing lesions, including presence of phlebitis and marked increase in IgG4⁺ cells, fulfilling the morphologic and immunohistologic criteria of IgG4-related sclerosing disease.^{5,6} The male predominance and occurrence in older adults are also characteristic of the latter condition. Furthermore, 2 patients showed additional manifestations of the disease, including IgG4-related chronic sclerosing sialadenitis of submandibular glands (both patients), parotid and orbital masses (common sites affected by the disease, although biopsy has not been obtained, in 1 patient), and lymphadenopathy (both patients).¹⁷ Although the systemic lymphadenopathy in case 3 could have been caused by lymphomatous involvement instead, the characteristic distribution of involved nodes (mediastinal, intraabdominal, and axillary) and dramatic response to steroid are more in keeping with lymphadenopathy associated with IgG4-related sclerosing disease.

Idiopathic cervical fibrosis (IgG4-related) can potentially be misdiagnosed as a lymphoma because of the worrisome infiltrative growth and dense lymphoid infiltrate. Awareness of this condition, lack of diffuse sheets of CD20⁺ B cells, and polytypia of the plasma cells would support a reactive process. The absence of conspicuous spindle cell proliferation and abundant IgG4⁺ plasma cells help distinguish this condition from fibromatosis and inflammatory myofibroblastic tumor. The lymphoid component is much more prominent than in analogous lesions of the mediastinum (mediastinal fibrosis) and retroperitoneum (retroperitoneal fibrosis), probably attributable to an earlier presentation of palpable mass lesions in the neck—sclerosis generally overtakes the lymphoid component with evolution of the lesion.⁶

An additional remarkable finding is that an extranodal marginal zone lymphoma has supervened in the background of IgG4-related cervical sclerosis in 1 case. A similar phenomenon has so far been reported to occur only in IgG4-related sclerosing disease of the ocular adnexa.^{4,23} Apparently the chronic inflammation provides the substrate for the emergence of the lymphoma, similar to the occurrence of extranodal marginal zone lymphoma in the acquired or autoimmune chronic inflammatory background in various mucosal sites.¹⁵ Of interest, this same patient also had classical Hodgkin lymphoma in a lymph node exhibiting features of IgG4-related lymphadenopathy, which is the first report of this

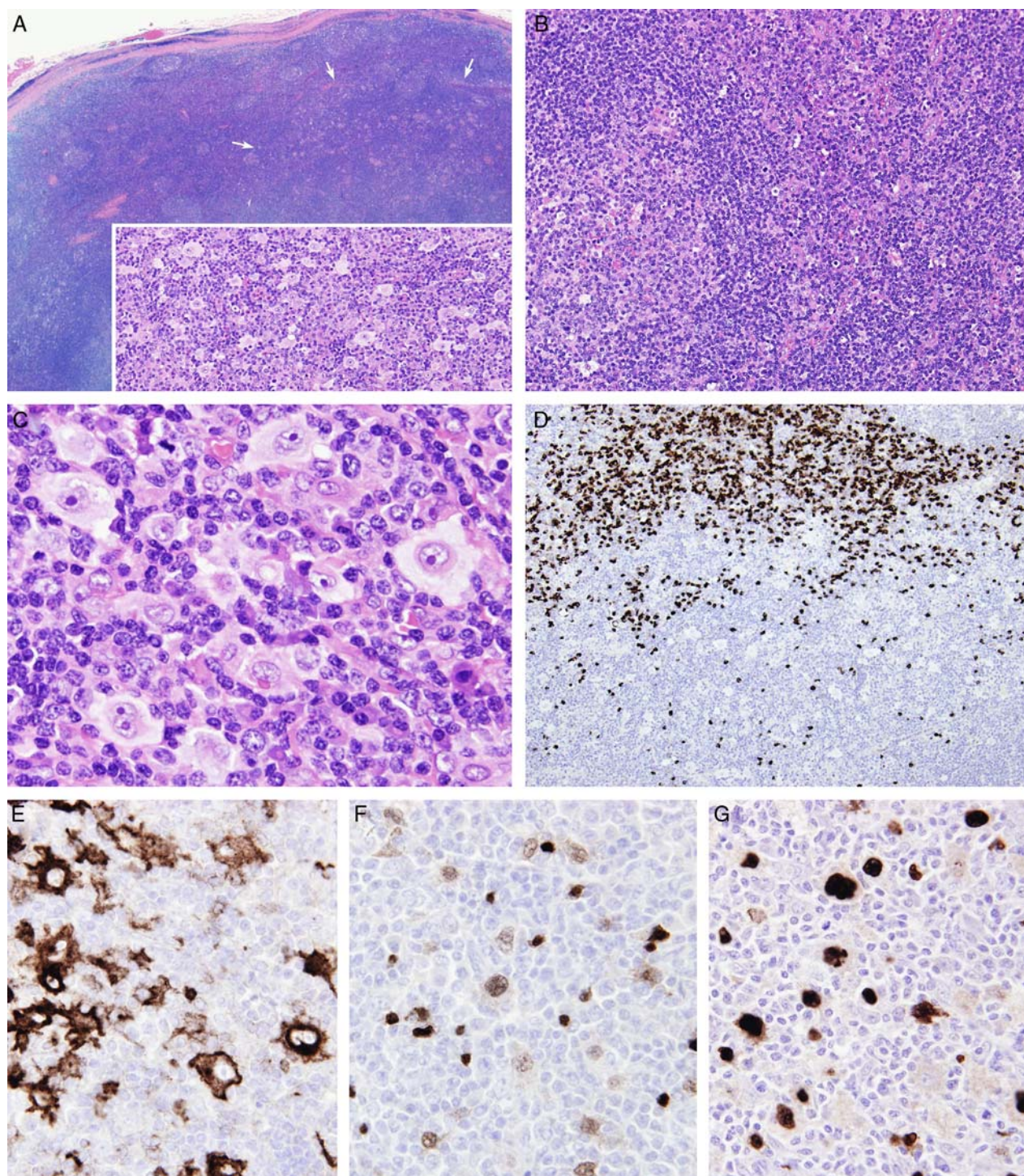


FIGURE 4. Lymph node of case 3. A, The lymph node shows reactive areas (left field) and abnormal foci (white arrows) with a mottled appearance (as shown in INSET). B, The reactive areas show lymphoid follicles separated by a plasma cell-rich interfollicular zone, compatible with IgG4-related lymphadenopathy. C, The abnormal foci show Reed-Sternberg cells and variants dispersed singly among small lymphocytes and plasma cells, compatible with classical Hodgkin lymphoma. D, Immunostaining for IgG4 shows numerous positive cells in the reactive areas (upper field), but few positive cells in the Hodgkin lymphoma component. E, The large neoplastic cells are CD30-positive. F, They show weak staining for PAX5. G, They show nuclear labeling for EBER on in situ hybridization. [full color online](#)

lymphoma type in such a setting. Unfortunately, immunoglobulin gene rearrangement cannot be shown in the Reed-Sternberg cells, precluding assessment of the clonal relationship with the extranodal marginal zone lymphoma. It is unlikely that the fibroinflammatory lesion in this particular case merely represents an unusual reaction to the lymphoma, because this patient has a long history of IgG4-related chronic sclerosing disease before the onset of lymphoma, and the latter does not seem to be the epicenter of the IgG4⁺ cellular infiltrate and sclerosing process of the lesion.

In a recent study from the Mayo Clinic following 111 IgG4-related sclerosing disease patients for a mean period of 3 years, 3 (2 with autoimmune pancreatitis and 1 with chronic sclerosing dacryoadenitis) developed lymphoma (2 diffuse large B-cell lymphomas or 1 B-cell lymphoma unspecified) in other sites, including the adrenal, liver, and kidney. The findings suggest an increased risk of lymphoma in general for these patients in comparison with the general population.²⁵

In conclusion, idiopathic cervical fibrosis represents a manifestation of IgG4-related sclerosing disease in the soft tissues of the neck, and may occasionally be complicated by lymphoma. Radical surgery is not required because residual disease or recurrence can be easily treated with steroid. However, at least a reasonable-sized biopsy has to be obtained to secure the diagnosis and to rule out other pathologies. Since this is a systemic disease, the patients require work-up and follow-up for the presence of additional sites of involvement.

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