Takayasu's Disease

A Case Report with an Angiocardiographic Study

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We have recently had the opportunity to study a young Japanese woman with so-called "Takayasu's" or "pulseless" disease. Although angiographic studies have been done previously,1, 2 we believe this case to be the first in which such clear-cut confirmation of the gross pathology of this condition has been demonstrated by this method.

Review

The original case report by Takayasu in 19083 in the medical literature of Japan noted only the eye findings, and later authors4-7 described the condition more completely. The condition is known by a number of terms, of which the most popular is pulseless disease. Takayasu's disease represents a prototype of the so-called "aortic arch syndromes," a term first used by Ross and McKusick. This latter term implies a number of different causative factors, and of the cases in the occidental medical literature, most are due either to syphilis, with its aortitis and aortic aneurysms, or to arteriosclerosis. In Japan, however, it is usually due to a peculiar form of panarteritis of the great vessels which has been studied most extensively by the Japanese. It appears to be a distinct entity, of yet unknown etiology, occurring largely in young females. Of the 122 cases reported in Japan, 111 were females and 11 were males. Sano and Shimizu, of Tokyo University, noted only 2 men in 36 hospitalized patients. This type of panarteritis has only rarely been noted in the rest of the world, but in view of some reports it does unquestionably occur.1, 2, 4, 5, 9-30 Other causes of "aortic arch syndrome" include chronic dissecting aneurysms, congenital abnormalities, upper mediastinal tumors, trauma, thrombophilia, and neurogenic and thoracic outlet syndromes. The etiology of idiopathic panarteritis of the great vessels in unknown. It may possibly fall into the spectrum of "collagen diseases."

The clinical features of this interesting disease are the consequence of obstructed blood flow to the cerebral, visual, and vascular systems. The most striking changes are of the vascular system and are noted in the peripheral pulses. These findings are noted in the main branches of the aortic arch; namely, the subclavian and carotid arteries and in their more peripheral branches. Though early in the disease there may be simply asymmetry of the pulse, there is eventual progression to a complete obliteration. Additional findings include thrills and bruits over the involved vessels prior to complete occlusion, differences or inability to measure

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the blood pressures in the arms, collateral thoracic circulation, and marked hypersensitivity of the carotid sinus in the more advanced stages of the disease. Hypertension of the lower extremities has been noted in some cases. Though trophic changes of the head and neck and sometimes perforated nasal septi occur, ischemic ulcers of the upper extremities have not been seen. Symptoms include cervical pain and tenderness over the vessels, fatigability, and claudication of the arms, shoulders, and jaws. Pain, coldness, weakness, and paresthesias of the arms, shoulders, and back have been noted.

The visual signs and symptoms develop with gradually diminished blood flow. The symptoms are flashes of light, field defects, altered visual acuity, visual disturbances on changing position, and eventually in the late stages, blindness. The findings in the retina are pallor and new vessel formation, especially around the optic papilla. Cataracts develop in the late stages. Optic atrophy, retinal atrophy and pigmentation, atrophy of the iris, microaneurysms, sluggish blood flow, decreased ocular pressure, enophthalmos, and superficial keratitis have been seen.

The cerebral symptoms are among the most important. They include aches, dizziness, syncope, vertigo, epileptiform seizures, transitory hemiplegias and hemiparesis, aphasia, mental and memory impairment, and emotional lability. They can be most troublesome on positional changes that reduce the cerebral blood flow. These may be due to an abnormally sensitive carotid sinus. Eventually death results from cerebral vascular insufficiency.

The single almost consistent laboratory finding is an elevated sedimentation rate. The Wassermann test is usually negative. In Japan a high percentage of tuberculin reactors has been noted but it is difficult to evaluate in view of the high incidence of tuberculosis in the population. Other laboratory studies except for the oscillometric studies show no consistent pattern.

As to prognosis, the course is quite variable and remitting and may be quite long. A number of cases have been followed for periods over 10 years. Ash-Upmark and Jervell have observed cases to 14 years. Pathologically, involvement of the aorta and of the vessels leading from the aortic
arch have been repeatedly demonstrated,9, 14, 19, 22, 26, 28, 29, 32 All or part of the thoracic and abdominal aorta may be involved, though affinity for the more proximal portions has been noted. The pulmonary artery has also been involved. The walls of the vessels are thickened, and extensive perivascular adhesions between the adventitia and surrounding tissues have been seen. The chronic inflammatory process involves the adventitia, media, and endothelium, and thrombus formation occurs with partial or complete obliteration of the lumens of supra-aortic and other branches of the aorta. Recanalization has also been seen. Round-cell infiltration involves all the layers of the vessel wall, and giant cells have also been noted in the inflammatory reaction. The more peripheral branch vessels are unremarkable until the disease is quite far advanced.

Case Report

One year prior to entry, the patient, I.P., a 19-year-old Japanese bride of an American serviceman, had noted severe generalized headaches associated with marked pain and tenderness of the left anterior cervical region. This had cleared spontaneously after several months, and no further symptoms were noted until September 1957. At that time, there was a recurrence of pain in the left side of the neck, and the patient was seen at the Tokyo Women's Medical College. The presence of an aneurysm of the left carotid artery was noted in an area of tenderness in the mid-neck region. A left carotid arteriogram (fig. 1) showed an aneurysm. The symptoms then partially cleared, but again recurred in late October, this time involving the right side of the neck. On November 2, 1957, the patient complained of severe right-sided headaches, nausea, and vomiting of 1 week's duration. An aneurysm of the base of the right common carotid artery just above the sternoclavicular joint was noted in the inflammatory reaction. The right carotid region similar findings were noted. The aneurysm was in the midcarotid region. The left subclavian pulse was weakly palpable and the right was full. The abdominal aorta was unremarkable, and both femoral and pedal pulses were full and symmetrical. The blood pressures in the legs were both 140/90. Both fundi were negative. The heart was not enlarged and the rhythm was regular at a rate of 84. No murmurs were noted.

Cardiolipid determinations were normal. A tuberculin test was negative. The corrected sedimentation rate was 36 and 42 mm, per hour, and the C-reactive protein was elevated at 5½ units. Total protein was 7.5 Gm. per cent, albumin 3.2 Gm. and globulin 4.3 Gm. per cent. Serum cholesterol was 297 and 286 mg. per cent on 2 occasions and the esters were 195 and 167 mg. per cent respectively. Multiple other studies were negative, including hemoglobin, hematocrit level,
white blood count, reticulocyte and platelet counts, bleeding and clotting times, clot retraction, prothrombin time, blood urea nitrogen and glucose, urinalysis, cephalin flocculation and thymol turbidity tests, van den Bergh test, alkaline phosphatase, serum glutamic acid transaminase level, cell preparations for lupus erythematosus disseminated, and electrocardiogram.

Roentgen studies showed clear lung fields (fig. 2). The heart was not enlarged; however, the upper mediastinum was widened. The aortic knob was markedly prominent and the aorta was slightly elongated. Fluoroscopy confirmed the widening and no pulsations were seen in this region. The aorta was knobby and the amplitude of the pulsations was noted to be damped. Laminography confirmed the widening of the upper mediastinum but no further information was obtained.

In view of the possibility of an arteriovenous fistula, it was decided to explore the right side of the neck. On November 12, operation disclosed evidence of arteritis and periarteritis. The common carotid artery was leathery, thickened, and in-

durated with an aneurysm at its base and stenosis in the midcarotid region from intravascular thrombus formation. A marked thrill, palpable over the stenotic area, disappeared on opening the carotid sheath. An arteriogram was attempted but was unsuccessful. In view of the marked inflammatory changes, no biopsy was done. A right carotid arteriogram (fig. 3) confirmed the presence of an aneurysm of the right carotid artery with stenosis both at the proximal region of the aneurysm and distally. The distal portion was shaggy and irregular, suggestive of intravascular thrombus formation. Angiocardiographic studies on November 22, 1957 (figs. 4-6), showed evidence of diffuse involvement of the aorta and its main brachiocephalic branches. The pulmonary arteries were unremarkable. The entire thoracic aorta exhibited alternating stenotic and dilated regions. Its main branches were of the double innominate pattern,33 and the left common carotid artery was poorly seen. The major cerebral flow appeared to be through the vertebral arteries. These vessels also showed irregularities in contour.
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Following discharge the patient again noted minimal pain and tenderness of the left carotid region. On a follow-up visit 1 month later she described back and shoulder pains but was otherwise unchanged. She came to the United States with her husband in January 1958.

DISCUSSION

The symptoms and findings of this type of “idiopathic panarteritis of the great vessels” are reviewed and a case of this condition in its early clinical stages is reported. The most interesting feature of this case is the angiocardiographic study and its correlation with the previously observed pathologic findings. The extensive involvement of the aorta and its brachiocephalic branches of the supra-aortic vessels was well demonstrated.

The picture of alternating stenosis and dilatation of the aorta and its branches indicated that despite the early clinical stage the vascular involvement was extensive. Intra-vascular thrombus formation was a prominent feature. No doubt the turbulent flow within the stenosed arteries brought about the multiple dilatations. The particular pattern of involvement demonstrated by this study should not be interpreted as the only pattern possible. Our study also reveals some interesting features of the dynamics of the circulation and of the compensatory mechanisms involved.1, 29, 34, 35 Cerebral flows occurred primarily through the vertebral arteries. These vessels showed some dilatation, perhaps due to the extra load caused by gradual obstruction of flow through the carotid arteries.

This form of “aortic arch syndrome” exhibits a characteristic clinical and pathologic picture. It is most common in young women, where symptoms and findings related to insufficient blood flow to the brain, eyes, and arms are usually accompanied by an elevated sedimentation rate. The pathologic features are quite distinctive.

SUMMARY

A case of “idiopathic panarteritis” (Takayasu’s disease) of the great vessels in a young Japanese woman is reported and the angiocardiographic studies are presented.

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ADDENDUM

Since the preparation of this paper an additional case has been reported in which an angiocardiographic study showed a very similar appearance.36

SUMMARIO IN INTERLINGUA

Es reportate un caso de “panarteritis idiopathie” (morbo de Takayasu) del vasos grande in un juvæne femina japonææ. Le correspondente studios angiocardiographic es presentate.

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