Zeitschrift: Bulletin der Schweizerischen Akademie der Medizinischen Wissenschaften = Bulletin de l'Académie suisse des sciences médicales = Bollettino dell' Accademia svizzera delle scienze mediche

Herausgeber: Schweizerische Akademie der Medizinischen Wissenschaften

Band: 2 (1946-1947)

Heft: 4

Artikel: The pathogenetic significance of the so-called endocarditis parientalis fibroplastica

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DOI: https://doi.org/10.5169/seals-306843

D. C. 616.126.002

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The Pathogenetic Significance of the so-called Endocarditis Parietalis Fibroplastica

By **W. Löffler**

Ladies and gentlemen,

In 1936 I described a peculiar form of endocarditis and I called this *disease*, or I would rather say this syndrome "endocarditis parietalis fibroplastica with eosinophilia of the blood".

This peculiar form of endocarditis which I have seen *twice only* in 30 years has been noted since by a series of investigators namely *Mumme*, *Büchler*, *Egger* (1944), *Smith* and *Furth*, *Böger*, *Roulet Basle* (1944) and others, a total, I believe, of 22 cases. The pathogenetic significance of this syndrom is greater than its frequency.

The *diagnostic-clinical* aspect of this condition is quite definite and clear, it is the aspect of the so-called *Fridl-Pick* type of general *central* stasis, with *this* important difference, that the *pericardium* in my cases proves to be *absolutely free*. On the other hand, the parietal *endocardium* of both ventricules was considerably thickened, the valves themselves remaining *free*. This thickening of the parietal endocardium amounts to 2,3 or even 4 mm. This thick and fibrous endocardium prevents the normal diastolic relaxation of the ventricules in a high degree. Besides this, more or less considerable thrombotic masses may diminish the capacity of the ventricules, filling them up to a certain extent. These conditions explain fully the *clinical aspect* of the disease, which is the same as in chronic constrictive pericarditis. It is a kind of *Fridl-Pick* condition in which not the *peri*cardium but the *endo*cardium prevents the normal diastolic relaxation of the ventricular myocardium.

The *eosinophilia* may be considered to be the expression of an allergic reaction rather than the result of the considerable circulatory stasis in the lungs.

As far as its symptomatology is concerned the endocarditis fibroplas-

tica with blood eosinophilia is a well defined clinical picture easily recognisable to those who have seen it. But this is not the subject of my communication.

Pathogenetically it is important to establish the relations existing between the endocarditis fibroplastica and other forms of endocarditis, such as endocarditis lenta, "*Felty*'s disease" and "*Still*'s disease", *Winiwarter-Bürger's* disease, (that is thrombo-angiitis obliterans).

The different cases of parietal endocarditis described up to now (cases of *Büchler*, *Deus*, *Löffler*, *Mumme*, *Roulet*) show this disease in rather *different stages of development* or if one may say so, *in different stages of recovery*. In all cases death occured through *mechanical* reasons. The condition of recovery has been the most advanced in the second case observed by myself. In this case the signs of local inflammation had almost completely disappeared and the formation of fibroid tissue, covering the endocardium was very advanced. (The patient did not die in the inflammatory stage but so to speak through the action of endocardial scars.)

In the case reported, the eosinophilia, as high as 70% in the beginning of the observation, that is 9 months before death, *de*creased to 9% two and a half months and to 2% a fortnight before death.

The *in*creasing cyanosis and dyspnœa during the last two months cannot therefore be the cause of the eosinophilia. The latter, decreasing with the development of the heart's failure is rather to be interpreted as a sign of a special allergic condition connected with the formation of fibrous tissue ; *it disappeared when this process had come to an end.* This was the case shortly before death, the fatal issue being merely mechanical.

Egger in a recent paper thinks that the eosinophilia is not an obligatory sign of the parietal endocarditis of the type in question. I think the eosinophilia is to be considered as a *phasic* sign. It depends on the completeness of the examinations, whether the sign is found or not. This peculiar character of the eosinophilia was emphasised just above. As long as the allergic condition in question is present, the eosinophilia is persistent. As soon as this phase is over, the eosinophilia drops. This is in striking analogy with the development of the fugacious pulmonary infiltration together with blood-eosinophilia (syndroma of *Löffler*). In those cases also eosinophilia is a phasic sign appearing and disappearing with the characteristic conditions according to the development of the pulmonary infiltration. If blood examinations are not made in sufficient number, one might overlook the eosinophilia, the blood-examinations being either too early or too late.

The clinical manifestation is the same as that of extended pericardial adhesions interfering with efficient contractions and dilatations of the right and left sides of the heart : venous engorgement, chronic passive congestion of the liver, as well as dyspnœa and pulmonary œdema i.e. chronic right *and* left sided cardiac failure.

No fever was observed during the evolution of the disease. On the other hand, the *possibility* of temperature-reactions existed, as the

patient showed a short fever-period 3 months before death when several pulmonary embolisme occurred. Bacteriæmia proved to be absent in as many as 9 examinations.

The case reported by *Deus*, which I had observed clinically in 1916 and which observation had led me to recognise the same *clinical* picture in 1936, died in an earlier stage of development than my last case. The eosinophilia was as high as 70,3% and *persistent*. Fever and bacteriæmia were *absent* during the whole clinical observation and streptoc. viridans was found only at the post mortem. In this case death occured in so early a stage that the fibrous transformation of the fibrine was not so far advanced as in the second case and the thrombotic process, indeed in regression, was still very marked.

In some cases of this peculiar form of parietal endocarditis (*Deus, Büchler*) streptococcus viridans or better "dissociated streptococci" could be found in the organs post mortem i.e. streptococci transformed from the more virulent to the less virulent types. The case of Deus and again the case of Roulet showed alterations in the kidneys (*Löhlein*'s, nephritis) which are considered to be fairly characteristic lesions in endocarditis lenta. These alterations were lacking in my last case. Marked eosinophilia occurs *very* rarely in the classical form of endocarditis lenta, though the eosinophiles are practically *never* lacking ; on the other hand bacteriæmia is, in endocarditis lenta, a *constant finding* throughout the whole disease. Bacteriæmia was as a rule absent in parietal endocarditis and was found only exceptionally.

We are therefore in the presence of a very peculiar form of endocarditis even apart from the localisation of the lesion (here on the wall-endocardium and there on the endocardium of the valves).

The parietal endocarditis can be considered as a condition *on its way to being cured* owing to a favourable allergic reaction of the body. This reaction, however, proves to be so strong as to produce scars to an extent which is *functionally fatal.* The organism kills itself mechanically when it has overcome the *toxic infectious* period. The classic form of endocarditis lenta shows a kind of equilibrium between the virulence of the streptococci and the defence is, by *itself, incurable*, as far as the infectious process is concerned. It can only be cured by a disturbance of this equilibrium by a *very* strong additional therapeutic action on the *cocci* (Penicillin). An increase of the immunological power would probably only lead to the formation of fibrous scars *on the valves* in analogy to the parietal endocarditis. In our case a further analogy with endocarditis lenta may be mentioned namely the existence of an old *valvular* lesion. The endocarditis lenta is known to be always a secondary disease,

preceeded either by endocarditis simplex with consequent valvular lesion or by a congenital malformation.

In my case the presence of an old mitral insufficiency and stenosis emphasised the *secondary* character of the parietal endocarditis. This secondary devolopment, however, did not touch the valves.

In the very clear and useful scheme of *Albertini* we would localize this disease near to the endocarditis lenta but decidedly verging towards the side of the rheumatic conditions i.e. comparatively *great resistance* rather than diminished virulence of the cocci.

In endocarditis lenta the needle of immunity oscillates uncertainly near the culminating point. It moves *decidedly towards the right*, that is towards scar-formation in parietal endocarditis, as is the case in thrombangiitis (*Buerger's* disease).

The eosinophilia of the blood is to be considered as an indicator of this peculiar condition. The scars, I think, are its patho-morphological expression. This constellation seems to me totally different from the phenomenon of recovery occuring under Penicillin-treatment, the action of Penicillin going *directly* against the cocci, eliminating them. In parietal endocarditis on the contrary, recovery is due rather to an increase of the immunisatory power of the body, which *step by step transforms the inflammatory lesion into fibrous tissue.*

Summary

Endocarditis parietalis fibroplastica with blood eosinophilia (*Löffler*) forms a clinically and pathologic-anatomically well-established syndrome of the rheumatic symptom complex. It is closest to the pathogenesis of Endocarditis lenta, but is distinguished from this latter form by the increased state of resistance of the organism (*Albertini*).

Zusammenfassung

Die «Endocarditis parietalis fibroplastica mit Bluteosinophilie» (*Löffler*) bildet ein klinisch und pathologisch-anatomisch wohl fundiertes Syndrom aus dem rheumatischen Formenkreis. Sie steht pathogenetisch der Endocarditis lenta am nächsten, zeichnet sich aber dieser gegenüber durch wesentlich erhöhte Resistenzlage des Organismus aus (*v. Albertini*).

Résumé

L'endocardite pariétale fibroplastique s'accompagnant d'éosinophilie sanguine (*Löffler*), forme un syndrome bien caraterisé du point de vue clinique et anatomo-pathologique, dans le domaine des formes rhumatis-

males. Sa pathogénie est la plus proche de celle de l'endocardite lente, mais s'en distingue par une résistance nettement plus marquée de l'organisme (*v. Albertini*).

Riassunto

L'Endocardite parietale fibroplastica accompagnata da eosinofilia (*Löffler*) costituisce una sindrome clinica e anatomo-patologica ben definita appartenente al gruppo delle affezioni reumatiche. Essa somiglia molto dal punto di vista delle patogenesi alla endocardite lenta, dalla quale si distingue però per una maggior resistenza da parte dell'organismo.

v. Albertini: Schweiz. Z. Path. u. Bakt. **1,** 163 (1938). – *Bäumler:* Dtsch. Arch. klin. Med. **1911,** 103. – *Böger:* Beitr. path. Anat. **81,** 441 (1929). – *Büchler:* Z. Klin. Med. **140,** 56 (1941). – *Deus, P.:* Korresp.bl. Schweiz. Ärzte **1916,** 1473. – *Egger, P.:* Schweiz. Z. Path. u. Bakt. **VII,** 237 (1944). – *Löffler, W.:* Schweiz. med. Wschr. **66,** 817 (1936). – *Mumme:* Z. Klin. Med. **138,** 22 (1940). – *Roulet, F.:* Schweiz. med. Wschr. **1944,** 427. – *Smith* and *Furth:* Arch. int. Med. (Am.) **71,** 602 (1943).